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4D Visions 2019 – Surgery without borders: Another landmark in the history of our society

Caros Sócios,

É com enorme orgulho que vos dou as boas vindas ao congresso SPCCTV 4DVisions 2019, sob o lema "Surgery without borders". Nesta edição, a quarta visão/dimensão é a da Sociedade Portuguesa de Cardiologia, uma das mais proeminentes sociedades cientificas nacionais.

Este congresso será, seguramente, um evento sem precedentes na história da Sociedade Portuguesa de Cirurgia Cardio-Torácica e Vascular (SPCCTV), face a uma combinação de circunstâncias inéditas:

- Pela colaboração da Sociedade Portuguesa de Cardiologia, dada a sua dimensão, capacidade de mobilização mediática e por ser um verdadeiro parceiro na vida real das especialidades que incorporam a SPCCTV;
- Pelo merecido destaque que será dado à especialidade de Cirurgia Torácica, que voltará a ter salas próprias para as suas comunicações e que, pela primeira vez na história da SPCCTV, a par com as restantes especialidades, contará com a entrega de um prémio consagrando o melhor trabalho na sua área – o Prémio Professor Doutor Esteves Pinto, honrando a escola Portuguesa de Cirurgia Torácica e tornando mais equitativa a relação das especialidades que compõem esta sociedade;
- Pela colaboração do Grupo de Estudos do Cancro do Pulmão na elaboração do programa científico de Cirurgia Torácica;
- Pela associação da 5ª edição do Thoracic Aorta Lisbon Symposium, ao congresso SPCCTV 4D Visions. Este prestigiado evento independente, organizado pelo Dr. Álvaro Laranjeira, vem aumentar a relevância e visibilidade do nosso congresso, permitindo alargar o leque de convidados nacionais e internacionais, que, de uma forma custo efectiva poderão participar em ambas as reuniões;
- E, finalmente, pela consolidação do modelo de congresso 4D Visions, criado em 2017, que tem demonstrado, pela sua modernidade e dinamismo, ser um sucesso.

Face a todas estas condições, estou confiante que teremos um painel de palestrantes e congressistas digno de nota, pelo que o congresso SPCCTV 4D Visions 2019 será um novo marco na história da SPCCTV.

Bem vindos ao congresso que celebra a multidisciplinaridade, porque juntos somos mais fortes!

Gonçalo Cabral | Vice-Presidente da SPCCTV

EDITORIAL

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Images in Clinical Medicine

The rapid progress of medical science and the invention of various medicines have benefited mankind and the whole civilization. Modern science also has been doing wonders in the surgical field. But, the proper and correct diagnosis of diseases is the primary necessity before the treatment. The more sophisticate the bio-instruments are, better diagnosis will be possible. The medical images play an important role in clinical diagnosis, teaching and researching. Medical imaging is often thought of as a way to represent anatomical structures of the body with the help of X-ray, computed tomography and magnetic resonance imaging. But often it is more useful for physiologic function rather than anatomy. With the growth of computer and image technology medical imaging has greatly influenced medical field. As the quality of medical imaging affects diagnosis the medical image processing has become a hotspot and the clinical applications wanting to store and retrieve images for future purpose needs some convenient process to store those images in details.

Your skill as a diagnostician is enhanced as your personal image bank grows and is committed to memory. Our image banks begin in medical school as we view pictures in lectures and textbooks, and they expand during our own clinical experiences. Studying and learning image patterns from any atlas - print or electronic - can enhance your expertise. Not all images, however, are retained and retrievable. Grotesque and disturbing images are retained because they are processed with strong emotional content. I contend that, in a similar way, images you photograph of patients who share their stories with you are likely to become memorable because of the highly personal context. These images will give you a wealth of material for self-instruction, teaching, and medical chart documentation.

If clinical photography is one of your interests, we encourage you to submit your best images and case descriptions for possible inclusion in this column. We are interested in clear, well-lit photographs accompanied by interesting stories that teach important practice principles. Whether or not you use photography in your own clinical work, we hope the detailed, informative cases in this SPCCTV Journal column will become an important resource for you in building your image bank.

Do you have images of compelling clinical cases of interest to cardiac, thoracic or vascular surgeons? We would like to publish them along with a brief description of the clinical presentation and a diagnostic question for readers. The case should include information on the differential diagnosis and treatment, the latter applying an evidence-based approach supported by current references. Multiple images may be submitted, and interesting connections between these images may be highlighted. See you soon…

Miguel Guerra | Editor-in-Chefe

EDITORIAL COMMENT

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Contemporary operative results of complete atriventricular septal defects

"If everyone is moving forward together, success takes care of itself."
Henry Ford

In a recent past, surgical correction of complete atrioventricular septal defects, carried significant morbidity and mortality related to residual intracardiac residual shunts, left atrioventricular valve regurgitation, pulmonary hypertension and cardiac arrhythmias. Operative mortality has been steadily decreasing from 10 to 15%[1,2], in the last decade of the 20th century to less than 3%[3] in present time, including no operative mortality in selected institutional series.[2,4]

The excellent surgical results reported in this paper from Sena et colleagues[5], highlight some of the advances that made possible this improvement, namely the mandatory echocardiographic control in the operative room, providing immediate quantification of the left atrioventricular valve regurgitation and presence of significant residual shunts, the advances on cardiopulmonary bypass and myocardial protection allowing for extra pump runs when necessary to achieve an optimal surgical result, the appearance of selective pulmonary vasodilators, specifically inhaled nitric oxide and oral sildenafil permitting the control of pulmonary hypertensive crisis, the overall improvement in the peri-operative care of these patients, namely in anaesthesia and pediatric intensive care allowing surgical repair in early infancy and low weight patients, thus avoiding the deleterious effect of the long-standing left to right shunt on ventricular performance and pulmonary circulation and, finally but not the least, the cumulative experience that cardiac surgeons in activity inherited from their less younger colleagues.

Comparisons of operative techniques are limited by different Eras, Surgeons, Institutions and variable anatomy. The three techniques have demonstrated similar good results respecting left AV valve regurgitation, left ventricular outflow tract obstruction and residual shunts, when tailored to each patient characteristics.

REFERENCES

1. Crawford FA, Stroud MR. Surgical repai of complete atrioventricularcseptal defect. Ann Thorac Surg.2001;72:1621-8.
2. Airaksinen R, Matila I, Jokinen E, Salminen J, Puntilla J et al. Complete Atrioventricular Septal Defect: Evolution of Results ina Single Center During 50 years. Ann Thorac Surg 2019; 107:1824-30.
3. T Louis JD, Jodkka U, Jacobs JP, Xia H. Contemporary outcomes of complete AVSD repair: Analysis of the STS congenital heart surgery database. Circulation 2010;122:S209-15.
4. Janai AR, Bellinghausen W, Turton E, Bevilacqua C, Zakhary W, Kostelka M, et al. Retrospective study of complete atrioventricular canal defects: Anesthetic and perioperative challenges. Ann Card Anesth. 2018; 21:15-21.
5. Sena A, Sheytanov V, Liebrich M, Narr A, Schweigmann U, Uhlemann F, Doll N, Tzanavaros I. Twelve years of complete atrioventricular septal defect repair. Rev Port Cir Cardiotorac Vasc. 2019; 26(3):187-193.

EDITORIAL COMMENT

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Rapid deployment prosthesis: a powerful tool to be used wisely

Rapid deployment (RD) valves have emerged in the surgical aortic valve replacement (SAVR) spectrum to facilitate the procedure. Traditional advantages attributed to these prostheses include shorter cardiopulmonary bypass (CPB) and cross-clamp times (XCT).[1] In 2019, there are two available: Intuity Elite™ (Edwards Lifesciences, Corp, Irvine, CA USA) RD prosthesis and Perceval™ (LivaNova PLC, London, UK) sutureless prosthesis. A third RD one, 3F Enable™ (Medtronic, Inc, Minneapolis, MN USA) is off the market since 2015.

According to the manufacturers, these devices are approved for aortic stenosis and steno-insufficiency patients. Typically, pure aortic regurgitation, endocarditis, multiple valve surgery and ascending aorta dilatation are contraindications for RD and sutureless valves. However, multiples reports have been published with successful off-label implantation of these devices in patients with concomitant mitral valve replacement,[2] supracoronary ascending aorta replacement,[3] bio-Bentall procedure,[4] and endocarditis.[5]

In fact, RD valves have some other advantages beyond better CPB and XCT times. They are useful in calcified aortic roots and are a valuable tool in minimally invasive procedures, mainly in right anterior minithoracotomy SAVR. Therefore, with increased experience and confidence from the surgical community, these expanding indications for RD valves will probably become even broader.

However, a word of caution must be written about this topic. In fact, the implantation technique for these valves seems to be easier than conventional stented valves. However, there are some particular aspects that require careful attention. For instance, a correct sizing is essential to achieve the best hemodynamic results. In fact, oversize is related to worse hemodynamic outcomes in sutureless valves, and perhaps a combination of CT-scan sizing and intra-operative sizing after native aortic valve removal would be more appropriate.[6] Additionally, these valves seem to be associated with increased pacemaker rate implantation and para-valvular leakage.[7] However, it is demonstrated that these complications can be mitigated by the learning curve overcome, which highlights the importance of dedicated teams with a significant RD valve SAVR caseload to improve the outcomes.[8]

Again, increased experience with these devices culminates into its broader application. Therefore, Santa Maria Hospital group must be congratulated for their expertise with RD valves, which is evident by this new application for them – combined SAVR with supra-coronary aortic replacement.[9] Their initial experience is very promising, showing outstanding CPB and XCT times. However, in this small sample, these reduced times do not appear to be associated with a significant improvement in clinical outcomes. Nonetheless, as this combined procedure with RD valves seems to be safe, it is essential to encourage their group and other Portuguese groups with significant experience with these devices to continuously pursuit innovation. In the end, there is no doubt that this will translate into better outcomes for our patients.

REFERENCES

1. Glauber M, Moten SC, Quaini E, et al. International Expert Consensus on Sutureless and Rapid Deployment Valves in Aortic Valve Replacement Using Minimally Invasive Approaches. Innovations (Phila). 2016 May-Jun;11(3):165-73.
2. Minh TH, Mazine A, Bouhout I, et al. Expanding the indication for sutureless aortic valve replacement to patients with mitral disease. J Thorac Cardiovasc Surg 2014;148:1354-9.
3. Concistrè G, Chiaramonti F, Bianchi G, et al. Aortic Valve Replacement With Perceval Bioprosthesis: Single-Center Experience With 617 Implants. Ann Thorac Surg. 2018 Jan;105(1):40-46.
4. Gennari M, Agrifoglio M, Polvani G. Composite graft using an Edwards Intuity Elite rapid deployment bioprosthesis for aortic root replacement. J Card Surg. 2017 Mar;32(3):193-195.
5. Ruchonnet EP, Roumy A, Rancati V, Kirsch M. Prosthetic Valve Endocarditis after Transcatheter Aortic Valve Implantation Complicated by Paravalvular Abscess and Treated by

Pericardial Patches and Sutureless Valve Replacement. Heart Surg Forum. 2019 Apr 4;22(2):E155-E158.
6. Cerillo AG, Amoretti F, Mariani M. Increased Gradients After Aortic Valve Replacement With the Perceval Valve: The Role of Oversizing. Ann Thorac Surg. 2018 Jul;106(1):121-128.
7. Ensminger S, Fujita B, Bauer T. Rapid Deployment Versus Conventional Bioprosthetic Valve Replacement for Aortic Stenosis. J Am Coll Cardiol. 2018 Apr 3;71(13):1417-1428.
8. Barnhart GR, Shrestha ML. Current Clinical Evidence on Rapid Deployment Aortic Valve Replacement: Sutureless Aortic Bioprostheses. Innovations (Phila). 2016 Jan-Feb;11(1):7-14.
9. Ferreira R, Junqueira N, Arruda R, Nobre A. Concomitant ascending aortic replacement and aortic valve replacement usingrapid deployment bioprosthesis. Rev Port Cir Cardiotorac Vasc. 2019; 26(3):195-198.

EDITORIAL COMMENT

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Cardiac surgery in Dialisys patients - can we do better than in the past?

Dialisys patients have always been considered difficult and high risk for cardiac surgery, due to extensive vascular disease, poor functional reserve, and frequently multiple cardiovascular lesions. Additional problems posed by these patients include difficult volume and eletrolyte control in the intra-operatory and post-operatory period, difficult hemostasis due to platelet disfunction, and susceptibility to infection.

In this regard, isolated experiences have been published all over the world, and risk scores have also included dialisys patients in their patient pool when being constructed. Still, even though cardiac surgery is somewhat frequently performed in theses patients, the outcomes are in some patients disappointing, while in other patients the intra-operatory and post-operatory period is almost as uneventful as in regular patients, albeit the programmed dialysis that always accompanies these patients.

Valvular percutaneous intervention has also opened new possibilities in these patients in whom cardiac surgery may be an excessive risk. Modern TAVI and mitral intervention techniques offer some of these patients a solution, but are far from being a treatment applicable to all - Biological valves (as all TAVI are) are susceptible to earlier structural valve deterioration in ESRD patients, mitral intervention is far from being long-lasting, and previous peripheral vascular procedures such as renal grafts in the iliac arteries and arm arteriovenous fistulas all complicate coronary and valvular percutaneous procedures. In addition, dialysis patients frequently present a coronary artery disease anatomy that is not amenable to percutaneous treatment (distal Left Main or ostial LAD disease, diffuse LAD disease, and very extensive calcification).

On the other hand, cardiac surgery frequently treats concomitant cardiac disease, associated valvular and coronary lesions are frequent in about 25% of cardiac surgery patients, and peripheral artery disease that may compromise saphenous harvest is very frequent in dialysis patients.

In this issue of SPCCTV, Ranchordás et al present their center´s experience with dialysis patients submitted to cardiac surgery (including 2 transapical TAVI´s). Their results are similar to the previous published experiences and reflect the good quality of Portuguese Cardiac Surgery. They also point out that higher EuroscoreII, CCS score and longer cross-clamp times (normally associated with concomitant procedures), are associated with a higher procedural/in-hospital mortality. Long term mortality is also higher in dialysis patients when compared to matched non dialysed patients.

So, a physician treating these patients may offer the normally very unsatisfactory optimal medical therapy, the total percutaneous option, the total surgical treatment or a hybrid modality. We should consider that these patients have a lower life expectancy than the non dialysed cardiac patients, even after controlling for the presence of cardiac disease (as Ranchordás et al have shown). The survival and MAACCE free advantage that surgery normally offers when compared with percutaneous therapy may be, in some cases, irrelevant, due to the progressive nature of end-stage renal disease.

Another important consideration that the paper by Ranchordás et al shows us is that, globally speaking, isolated open procedures in these patients are safe and should be offered liberally (with the probable exception of very frail patients), with the same constraints that the surgery in the general population has. Multiple vessel coronary disease in the presence of peripheral arterial disease may occasionally be treated by an hybrid approach to avoid risky saphenous vein harvesting.

Concomitant procedures, like aortocoronary surgery, is not specifically contra-indicated, but longer cross--clamp times may guide the surgeon to opt for a hybrid approach, reserving the lesions less amenable for percutaneous treatment for the surgeon to treat. Surgical strategies to avoid longer cross clamp times, like performing coronary grafting on the beating heart, or using rapid--deployment aortic valves, may also help diminish cross clamp times (if this has an impact on survival remains to be determined).

Dialysis patients should be, and will continue to be offered the most definitive treatment for cardiac lesions. Surgeons and Cardiologists should also continue to strive for better outcomes and better long term results, and it is in difficult and risky patients that the Heart Team will probably more positively influence the outcomes.

REFERENCES

1. Ranchordas S, Madeira M, Pereira T, Branco P, Gaspar A, Marques M, Calquinha J, Abecasis M, Neves JP. Cardiac surgery in patients with dialysis-dependent end stage renal failure single centre experience. Rev Port Cir Cardiotorac Vasc. 2019; 26(3):199-204.
2. Deutsch O, Spiliopoulos K, Kiask T, Katsari E, Rippinger N, Eichinger W, Gansera B: Cardiac surgery in dialysis-dependent patients: impact of gender on early outcome in single-center experience with 204 consecutive cases. Thorac Cardiovasc Surg 2013, 61(1):22-28.
3. Bäck C, Hornum M, Møller CJH, Olsen PS. Cardiac surgery in patients with end-stage renal disease on dialysis. Scand Cardiovasc J. 2017 Dec;51(6):334-338. doi: 10.1080/14017431.2017.1384565. Epub 2017 Oct 4.

EDITORIAL COMMENT

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Carbon dioxide production during cardiopulmonary bypass: continuous measure and clinical relevance

Carbon dioxide production during cardiopulmonary bypass derives from both the aerobic metabolism and the buffering of lactic acid produced by tissues under anaerobic conditions. Therefore, carbon dioxide removal monitoring is an important measure of the adequacy of perfusion and oxygen delivery. However, routine continuous monitoring of carbon dioxide removal is not widely applied.

Carbon dioxide (CO_2) removal monitoring during cardiopulmonary bypass (CPB) is considered a recommended guideline for practice by the American Society of Extracorporeal Technology and a standard of practice by the Australian New Zealand College of Perfusion.[1,2] This monitoring is usually performed through capnometric analysis of gases from the exhaust port of the oxygenator. Despite its relative easiness, this practice is not routinely applied.

CO_2 tension (PCO_2) as measured by capnometry is representative of the complex interaction of CO_2 production (VCO_2) by cells and its elimination by the natural or artificial lung. Therefore, this technique may provide important information, not only on the efficacy of CO_2 removal, but even on the metabolic status of the peripheral organs and on the adequacy of their perfusion with respect to their oxygen (O_2) needs.[3]

The hollow-fibre/membrane oxygenators commonly used on CPB have totally different performance in terms of CO_2 clearance with respect to the natural lung. Actually, the ventilation/perfusion ratio of the natural lung is around 0.8 (higher at the top and lower at the bottom of the lung in the orthostatic position). An increase of this ratio, as happens in the case of low pulmonary blood flow, creates the conditions for a $PvCO_2 - ePCO_2$ gradient (Mixed venous carbon dioxide tension - Exhaled carbon dioxide tension). Conversely, artificial lungs have a much higher efficiency in clearing venous blood from the CO_2, with an optimal ventilation/perfusion ratio of around 0.4-0.5.[4]

This property of the artificial lung offers a great advantage for the measurement of VCO_2 during CPB. The ability of the oxygenator to clear off the CO_2, even at a low ventilation/perfusion ratio, avoids the onset of a $PvCO_2 - ePCO_2$ gradient and the $ePCO_2$ measured at the exhaust port of the oxygenator can be reliably used to assess the VCO_2.

Infra-red spectography is the most popular means currently used to monitor CO_2 and most centres that use oxygenator exhaust capnography routinely use sidestream devices because of their ease of use.

Initially, capnography was mainly used to measure metabolic CO_2 production and to estimate $PaCO_2$. Many studies show a good correlation between ePCO2 and uncorrected $PaCO_2$ during clinical cases,[5-7] making it easier to maintain a specific target $PaCO_2$ during CPB. Although there is a good linear correlation between $ePCO_2$ and uncorrected $PaCO_2$, there is some deviation, especially during the rewarming phase. This deviation is the same for oxygenators of the same type and brand, but can differ between types and brands.

More recent research showed that VCO_2 is a good predictor of anaerobic metabolism[8] and, as such, can help to reduce CPB-related morbidity. Indeed, CO_2-derived parameters are more rapid and sensitive than O2-derived parameters in detecting anaerobic metabolism.[9] A combination of DO_2, VO_2 and VCO_2 parameters was significantly associated with the risk of postoperative renal insufficiency.[10]

The potential clinical relevance of VCO_2 monitoring goes far beyond the safety control of the maintenance of an adequate $PaCO_2$ of the patient throughout the CPB procedure. Certainly, the on-line measurement of $ePCO_2$ at the exhaust port of the oxygenator may prompt sweep gas adjustments to rapidly adjust the systemic $PaCO_2$ during the different phases of CPB. Increased values of $ePCO_2$ as an expression of increased VCO_2 can be found at the release of the cross-clamp, due to the reperfusion of the heart (anaerobic CO_2) or during the rewarming phases after deep hypothermia, as an expression of the increasing VO_2 (aerobic CO_2) and of the decreased solubility of CO_2. This last mechanism should be considered when assessing

VCO_2 during CPB cooling and rewarming phases, where the changes in CO_2 solubility respectively decrease and increase the $ePCO_2$.

Nowadays, the routine use of low temperatures on CPB has been replaced by moderate hypothermia or normothermia in many institutions. However, CPB temperatures $<28°C$ may still be used in congenital heart surgery and for specific interventions of high complexity. Within the setting of profound hypothermia, the changes in CO_2 solubility result in corresponding changes in pH, with a reflection on cerebral blood flow. To compensate for the low values of CO_2 at low temperatures, the pH- stat strategy considers the addition of exogenous CO_2 to the sweep gas.

Another exogenous source of CO_2 may come from the flooding of the surgical field to prevent the formation of large air bubbles inside the heart chambers. The finding of elevated values of $ePCO_2$ is rarely attributable to a failure of the oxygenator.

Apart from this, the measurement of the VCO_2 may offer important information on the adequacy of the perfusion in terms of DO_2 (pump flow x arterial O_2 content). In the pathophysiology of CO_2 production, under conditions of inadequate DO_2, there is an excess CO_2 production as a result of lactic acid buffering. Therefore, VCO_2 may be considered as an indirect marker of lactate increase.

The use of DO_2 and VCO_2 to guide the perfusion management are now included in the concept of the "Goal Directed Perfusion (GDP)".[12-14] The GDP concept considers that the goal of perfusion is to maintain an adequate oxygen supply to all the organs, avoiding the patient entering into the anaerobic zone.

Despite the many possible applications of CO_2-derived parameters during CPB, very few studies have been published in this area. The expanding concept of GDP will probably increase the interest of clinicians and researchers in these measurements. Further studies on the clinical relevance of CO_2 production monitoring are warranted.

The article from Valdir Filho[15] has revived this subject. Shows an available technique to assess carbon dioxide production and removal and the clinically relevant applications of carbon dioxide-related variables as markers of the adequacy of perfusion during cardiopulmonary bypass.

REFERENCES

1. Baker RA, Bronson SL, Dickinson TA, et al. Report from AmSECT's International Consortium for Evidence- Based Perfusion: American Society of Extracorporeal Technology Standards and Guidelines for Perfusion Practice: 2013. J Extra Corpor Technol 2013; 45: 156–166.
2. The Australian and New Zealand College of Perfusion. Regulations and Guidelines for Perfusionists. Available at: http://esvc000803.wic050u.server-web.com/Documents/ANZCP%20Regulations.pdf. Accessed December 17, 2015.
3. Wasserman K, Whipp BJ, Casaburi R. Respiratory con- trol during exercise. In: NS Cherniack, JG Widdicombe (Eds), Handbook of Physiology, Section 3: The Respiratory System. American Physiological Society, Bethesda 1986, pp 595–619.
4. Pybus DA, Lyon M, Hamilton J, Henderson M. Measuring the efficiency of an artificial lung: 1. Carbon dioxide transfer. Anaesth Intens Care 1991; 19: 421–443.
5. Alston RP, McNicol J. Oxygenator exhaust capnography: an in vitro evaluation. J Cardiothorac Anesth 1988; 2: 798–802.
6. Zia M, Davies FW, Alston RP, Anaes FC. Oxygenator exhaust capnography: a method of estimating arterial carbon dioxide tension during cardiopulmonary bypass. J Cardiothorac Vasc Anesth 1992; 6: 42–45.
7. Weightman WM, Sheminant MR. Oxygenator exhaust capnography as an index of arterial carbon dioxide ten- sion during cardiopulmonary bypass using a membrane oxygenator. Br J Anaesth 2000; 84: 536–537.
8. Ranucci M, Isgrò G, Romitti F, Mele S, Biagioli B, Giomarelli P. Anaerobic metabolism during cardiopulmonary bypass: predictive value of carbon dioxide derived parameters. Ann Thorac Surg 2006; 81: 2189 –2195
9. Mekontso-Dessap A, Castelain V, Anguel N, et al. Combination of venoarterial PCO2 difference with arteriovenous O2 content difference to detect anaerobic metabolism in patients. Intensive Care Med 2002; 28: 272–277.
10. de Somer F, Mulholland JW, Bryan MR, Aloisio T, Van Nooten GJ, Ranucci M. O2 delivery and CO2 production during cardiopulmonary bypass as determinants of acute kidney injury: time for a goal-directed perfusion man- agement? Crit Care 2011; 15: R192.
11. Dres M, Monnet X, Teboul J-L. Hemodynamic manage- ment of cardiovascular failure by using PCO2 venous- arterial difference. J Clin Monit Comput 2012; 26: 367–374.
12. Dijoy L, Dean JS, Bistrick C, Sistino JJ. The history of goal--directed therapy and relevance to cardiopulmonary bypass. J Extra Corpor Technol 2015; 47: 90–94.
13. Ranucci M, Aloisio T, Carboni G, et al. Acute kid- ney injury and hemodilution during cardiopulmonary bypass: a changing scenario. Ann Thorac Surg 2015; 100: 95–100.
14. Ranucci M, Romitti F, Isgrò G, et al. Oxygen delivery during cardiopulmonary bypass and acute renal failure after coronary operations. Ann Thorac Surg 2005; 80: 2213–2220.
15. Assis-dos-Reis-Filho V, Lopes-de-Oliveira E, Scramim JF, Sanga MA, Arrais-dos-Santos M. Benefits of continuous monitoring of PCO2 obtained from a system applied to membrane oxygenator exhaustion of the cardiopulmonary bypass circuit. Rev Port Cir Cardiotorac Vasc. 2019; 26(3):205-208.

EDITORIAL COMMENT

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Lung hernia: A rare consequence of a chest trauma

It is not possible to speak of lung hernia (LH), without referring to its main cause which is chest trauma, the kinetics involved and it's results that lead to the development of a hernia.

Significant thoracic Trauma (TT) is not as uncommon as one might think. All over the country, patients with traumatic chest wall injuries show up daily at emergency departments of peripheral and central hospitals. The spectrum runs from the major trauma to the minor one, affecting only the chest cavity, with a wide range of causes, some quite unusual.

The initial approach of a patient with a TT, includes the evaluation of chest wall integrity as well as haemodynamic and respiratory dynamics, with the objective of defining priorities in treatment.

All doctors involved in trauma care, from the pre-hospital emergency to the Intensive Care Unit, should be trained in the initial evaluation of a TT. These patients are not necessarily Thoracic Surgery patients, as specialists are seldomly present in peripheral hospitals, but consultation should be requested whenever needed.

TT treatment is multidisciplinary in most case scenarios, with active cooperation of medical and surgical specialists, as the patients' needs so determine.

Lung hernia or intercostal lung hernia are synonyms of the same rare pathological entity.[1,2,3,4,5]

For a LH to develop, lung parenquima needs to protrude through a gap in the thoracic wall, by loss of integrity of the costal grid which can have a congenital cause due to a defect in the sibson's fascia or be acquired.[1,2]

Aquired LH occur as compications of TT, or as a complication of thoracic surgery, although spontaneaous development does occur in 30% or associated to local pathological transformation.[2]

We know that acquired LH has no direct relation to the mechanism of trauma or its kinetic, for it can be due to large or small chest wall trauma. LH can be diagnosed immediately after trauma, weeks or even years after the traumatic event.[3]

Early diagnosis, highly depend on the consequences on the chest wall and chest cavity, and the symptoms at presentation. Late diagnosis most often happens in asymptomatic individuals as imaging findings when investigating other complaints.

In both cases, ethiological investigation should reveal its cause, as is the case of the article by Sara Lopes et al., published in this number of Revista Portuguesa de Cirurgia Cárdio-Torácica e Vascular,[4] on the case of a LH cause by an unusual chest wall trauma, occurring 14 years before.

Due to it rarity, this pathology is seldomly described in literature, with the publication of around 300 cases, mostly as isolated case reports.[1,5] Approximately 20% of the publications report congenital hernias and 80% of acquired defects.[2]

The mechanics of the original trauma is unusual, even in a Country where such cultural events still attract large number of people. The trauma victim was assisted in a Hospital setting and deemed for nonsurgical management of his chest wall trauma. Only after 14 years, when his COPD progressed, was the LH diagnosed, identified on inspection, asymptomatic. After being evaluated by a thoracic surgeon, it was decided not to correct it.

When this pathology has no symptoms, it is usualy not operated, but it can be corrected at the patients request, if a visible bulging in the thoracic wall makes him unconfortable.

These cases can and should be refered to a thoracic surgery outpatient clinic for evaluation. On rare occasions cooperation with plastic surgery may be required.

After the initial trauma, follow-up should be maintained, and the role of respiratory rehabilitation cannot be underestimated in the infirmary and after discharge, reducing permanent disabilities and promoting an earlier return to active life.

There are no publications to support high level scientific recommendations for the treatment of LH.[2] The treatment remains dependent on the experience and common sense of the multidisciplinary team treating these lesions.

REFERENCES

1. Bikhchandani J, Balters MW, Sugimoto JT. Conservative management of traumatic lung hernia. The Annals of thoracic surgery. 2012;93(3):992-4. Epub 2012/03/01.
2. Sílvia Raquel Coelho da Silva, e Col.: Hérnia pulmonar intercostal encarcerada em contexto de trauma toraco-abdominal. Relato de caso e revisão da literatura. Rev. Port. Cir. no.41 Lisboa jun. 2017.
3. John C Maybery MD et al: The fractured rib in chest wall trauma. Chest Surg.Clinic North Am. Vol.7 nº2:239-261 May 1997.
4. Lopes S, Costa R, Maciel J, Casanova J, Bastos PC, Pinho P. Lung hernia related with a rope bullfight case report. Rev Port Cir Cardiotorac Vasc. 2019; 26(3):219-222.
5. David JS, Tassin C, Maury JM. Posttraumatic pulmonar hérnia. Thorax. 2013;68(10):982.

ARTIGO ORIGINAL ORIGINAL ARTICLE

TWELVE YEARS OF COMPLETE ATRIOVENTRICULAR SEPTAL DEFECT REPAIR

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Abstract

Background: Surgical repair is the standard treatment for complete atrioventricular septal defect. At our institution, this repair is performed by single patch, modified single patch or two patch techniques, according to the surgeon preferences and the surgical anatomy of the defect. The goal of this study was to evaluate our results from the last twelve years.

Methods: From June 2006 to June 2018, 81 children with complete atrioventricular septal defect (without tetralogy of Fallot or unbalanced ventricles) were submitted to surgical repair at our institution. Data from all patients was retrospectively collected and evaluated.

Results: The average age was 6.9 ± 13.7 months and 84% had Down syndrome. Eighty percent were symptomatic and 6 patients were previously submitted to pulmonary artery banding. No more that mild left atrioventricular valve insufficiency was found in 84% and 89% of the patients, at discharge and follow-up, respectively. Small residual septal defects were present in 27% at discharge; during follow-up, 41% of these closed spontaneously. Pulmonary hypertension at discharge and follow-up appeared in 3.7% and 1.3%, respectively. Permanente pacemaker was implanted in 3 patients. Left ventricle outflow tract obstruction was found in 3 patients and 2 needed surgical correction. At follow-up (40 ± 38 months), 90% of the patients presented NYHA functional class I. No significant differences in the main repair outcomes were found between techniques, with the exception of small residual septal defects, although the groups were unmatched.

Conclusions: Overall and regardless of the technique used for the repair of complete AVSD, good early and midterm outcomes were achieved.

INTRODUCTION

Atrioventricular septal defects (AVSD) are a group of common congenital heart defects with an *ostium primum* defect and an (separated or common) atrioventricular (AV) orifice; an inlet ventricular septal defect may or not be present.[1] According to the ventricular defect size and the status of the AV valves, it can be classified as complete, intermediate or partial. The complete type presents with two components, one ventricular (ventricular septal defect - VSD) and another atrial (atrial septal defect - ASD), and a single common AV valve. In addition, other heart anomalies can coexist, sometimes as part of syndromes, most frequently, Down syndrome.[2]

If untreated, 50% of the patients with complete AVSD die in the first year of life due to heart failure and pulmonary infections. Untreated patients develop pulmonary hypertension (PHT) and Eisenmenger syndrome, therefore, an early surgical intervention is necessary.[3] However, the surgical treatment is challenging, requiring closure of the septal defects and repair of any AV valves dysfunction. Currently, 3 different techniques are commonly used: single patch technique (that corrects the septal defect with one single patch, dividing the bridging leaflets and then reattaching them to the patch)[4]; two-patch technique (that uses separated patches to correct de ASD and VSD, fixing the valve leaflets between them)[5]; and modified single patch technique or "Australian" technique (where the VSD is directly attached to the valve leaflets and the ASD closed with a patch).[6] So far, none have shown superiority over the others.[7,8]

The classic single patch technique was the method of first choice in our initial experience, with the modified single patch technique used by favorable anatomy. In 2013 the two-patch technique was added to the routine.

The aim of this study was to review the results of our experience using these 3 techniques to repair complete AVSD over the last 12 years.

PATIENTS AND METHODS

Population and Methods

Between June 2006 and June 2018, 81 children with complete AVSD, without tetralogy of Fallot or unbalanced ventricles, were submitted to surgical repair at our center. Single-patch technique was used in 41 patients; modified single patch technique and two-patch techniques were used in 20 patients each. The clinical data of all patients was retrospectively reviewed, with respect to patients' preoperative characteristics, operative parameters, perioperative outcomes and follow up.

Surgical aspects

Operations were performed after median sternotomy, on cardio-pulmonary bypass with bicaval cannulation (in mild to moderate hypothermia). Cardioplegic arrest was achieved with crystalloid cardioplegia. Intraoperative echocardiography was routinely performed in each patient; suboptimal results - severe AV regurgitation; moderate/severe AV or left ventricle outflow tract [LVOT] obstruction stenosis; residual ASD/VSD> 2 mm - were revised.

Statistical analysis

Statistical analysis was carried out using SPSS v24. Continuous variables were treated as mean and standard deviation and compared with t-student and ANOVA tests. Categorical variables were summarized as the number and/or percentage of subjects in each category and compared with Chi square/Fisher's exact testes.

RESULTS

Baseline

Overall, the patients were 6.9 ± 13.7 months old, 52% were female, the mean weight was 5.2 ± 2.9 kg, 84% had Down syndrome, 69% had another atrial septum defect, and 7.4% were previously submitted to PA banding. Eighty percent were symptomatic.

Baseline characteristics for each technique are summarized in table 1. The group of patients treated with the modified single patch technique (mSPG) were older on average, had more body weight, more Rastelli type A defects, less cases of Down syndrome, lower frequency of PHT and no previous PA banding. The groups of patients treated with the single patch (SPG) and two patch (TPG) techniques were more similar in the baseline characteristics.

Surgical Procedure

Surgical times varied with concomitant procedures and reoperation. The mean surgical times of the patients treated with a simple combination of procedures (AVSD and left AV valve valve repair, with/without persistent ductus arteriosus ligation) is described in the Table 2.

Closure of preexisting PFO/ASD II was the most common additional procedure in all groups (Table 3).

Intraoperative revision occurred less in the SPG (4.8%); TPG and mSPG had similar number of revisions (25% and 20%, respectively). Left or right AV-valve repair was the most frequent reason for revision.

Table 1 **Base characteristics of the patients by technique**

	Two Patch n=20	Single Patch n=41	Modified Single Patch n=20	*p*
Sex (female/male)	50/50 %	49/51 %	50/45 %	0.945
Age (months)	5.4 ± 3.7	6.4 ± 16.0	9.3 ± 15.2	0.636
Weight (kg)	5.2 ± 1.2	4.6 ± 3.2	6.3 ± 3.1	0.083
Height (cm)	60.4 ± 6.6	56.0 ± 12.9	54.5 ± 13.1	0.032
Rastelli type (A, B, C)	45 | 5 | 50 %	19 | 5 | 76 %	60 | 0 | 35 %	0.011
NYHA class (I, II, III, IV)	30 | 25 | 25 | 20 %	12 | 42 | 44 | 2 %	25 | 55 | 20 | 0 %	0.014
Comorbidity				
Down syndrome	95 %	85 %	70 %	0.028
PFO or ASD type II	85 %	59 %	75 %	0.089
Hypothyroidism	30 %	24 %	25 %	0.890
Former premature baby	15 %	7 %	5 %	0.484
Previous PA banding	20 %	5 %	0 %	0.037
Patent ductus arteriosus	10 %	5 %	20 %	0.178
Left superior vena cava	10 %	7 %	5 %	0.833
Pulmonary hypertension	70 %	78 %	45 %	0.034

ASD - Atrial septal defect; NYHA - New York Heart Association; PA - Pulmonary artery; PFO - patent foramen ovale; RBBB - Right bundle branch block
Continuous variables are presented in the format mean ± standard deviation (minimum - maximum values).

Table 2 **Surgical times**

	Single Patch n=29	Modified Single Patch n=15	*p*
Aorta Clamping (minutes)	119 ± 21.1 (79)	85 ± 18.4 (60)	101 ± 14.8 (77)
Cardiopulmonary bypass (minutes)	179 ± 31.1 (108)	128 ± 30.1 (87)	150 ± 32.1 (113)
Operative time (minutes)	261 ± 34.8 (192)	213 ± 30.4 (160)	229 ± 32.3 (180)

Only cases were atrioventricular septal defect repair plus left atrioventricular valve repair plus interatrial communication closure with or without ductus arteriosus ligation were performed are included in this table. The groups were unmatched.
Continuous variables are presented in the format mean ± standard deviation (minimum value).

Table 3 **Surgical procedures**

	Single Patch n=41	Modified Single Patch n=20	Two Patch n=20	*p**
Additional procedures	-	-	-	-
Right AV valve repair	6 (15%)	1 (5%)	0	0.129
PFO/ASD II correction	22 (54%)	15 (75%)	17 (85%)	0.034
by direct suture	14 (34%)	13 (65%)	11 (55%)	-
with pericardium patch	8 (20%)	2 (10%)	6 (30%)	-
Ductus arteriosus ligation	2 (4.8%)	5 (25%)	2 (10%)	0.063
PA de-banding (± augment plasty)	2 (4.8%)	0	4 (20%)	0.037
Intra operative revision	2 (4.8%)	4 (20%)	5 (25%)	0.318
Left AV valve insufficiency	1 (2.4%)	2 (10%)	4 (20%)	-
Right AV valve insufficiency	0	2 (10%)	0	-
VSD	1 (2.4%)	0	(1)	-
PA stenosis	0	0	1 (5%)	-

ASD - Atrial septal defect; AV - Atrioventricular; AVSD - Atrioventricular septal defect; PA - Pulmonary artery; PFO - Patent foramen ovale; VSD - Ventricular septal defect.
*The groups were unmatched.

ICU course

The mSPG had a shorter ventilation (4.1 ± 3.0 days), needed less catecolamines (4.6 ± 3.0 days) and phosphodiasterase inhibitors (2.3 ± 2.4 days) support and a shorter ICU (8.7 ± 5.4 days) and in-hospital (17.4 ± 7.5 days) stay (Table 4), whereas the amount of catecholamines and PDI didn't reach statistical significance.

Despite considerable numeric differences in the studied complications, none reached statistical significance, with urinary infection with identified agent being the exception (see Table 4 for details). There was no wound infection in the postoperative period.

Atrioventricular valves

Overall, 77% of the patients presented with a mild or trivial left AV valve insufficiency preoperatively. No more than a mild regurgitation was found in 84% at discharge and in 89% at follow-up; four patients (3 in the SPG and 1 in the TPG) needed to be reoperated in the early postoperative period due to severe left AV valve insufficiency. Freedom from moderate to severe regurgitation preoperative, postoperative and at follow-up were, 76%, 85% and 90% in the TPG, 78%, 88% and 85% in the SPG, and 75%, 75% and 90% in the mSPG, respectively (Figure 1A).

For the right AV valve, not more than a mild insufficiency was present in 95% of the patients at discharge and 96% at follow-up. Groups details are represented in Figure 1B.

There was no relevant (more than mild) stenosis of either the right or left AV valves, both early postoperative or during follow up.

None of the groups reached statistical significance, regarding the left and right AV valves insufficiency, in the in-hospital and follow-up phases.

Residual septal defects

During in-hospital stay, 1 patient in the SPG (2.4%) needed reoperation due to a large residual VSD; in the TPG, 1 patient was reoperated because of severe left AV valve insufficiency (as mentioned above) and, concomitantly, the VSD was also revised because of residual large VSD. In both cases, the VSD could be almost completely closed. At discharge, 45% of the patients in the TPG presented with small residual septal defects (ASD or VSD) (<2mm); the incidence was 35% in the mSPG and 17% in

Table 4 **Early post-operative results**

	Single Patch n=41	Modified Single Patch n=20	Two Patch n=20	*p**
Ventilation (days)	7.1 ± 3.8	4.1 ± 3.0	6.9 ± 4.0	0.011
Catecolamines (days)	5.9 ± 3.1	4.6 ± 3.0	5.9 ± 3.7	0.427
Phosphodiasterase inhibitors (days)	6.4 ± 8.1	2.3 ± 2.4	5.3 ± 4.1	0.113
ICU stay (days)	14.5 ± 11.5	8.7 ± 5.4	17.8 ± 11.8	0.026
Hospital stay (days)	26.5 ± 15.4	17.4 ± 7.5	31.9 ± 16.3	0.006
Post-operative complications Suspected infection (at least one) [confirmed with identified agent]	71 % [27 %]	45 % [10 %]	70 % [60 %]	0.119 [0.002]
Suspected infection (at least one) [confirmed with identified agent]	12 % [10 %]	10 % [10 %]	35 % [35 %]	0.053 [0.029]
Suspected urinary infection [confirmed with identified agent]	34 % [17 %]	30 % [5 %]	40 % [20 %]	0.799 [0.347]
Suspected respiratory infection [confirmed with identified agent]	2 % [2 %]	0% [0 %]	5 % [5 %]	0.595
Pulmonary hypertension crisis	17 %	20 %	45 %	0.051
Arrhythmia**	32 %	15 %	30 %	0.367
Permanent pacemaker implantation	2.4 %	5 %	5 %	0.830
Chylothorax	17 %	15 %	30 %	0.407
Atelectasis or Pneumothorax (significant)	20 %	20 %	30 %	0.628
Death	0 %	0 %	0 %	-

ASD - Atrial septal defect; AV - Atrioventricular; AVSD - Atrioventricular septal defect; PA - Pulmonary artery; PFO - Patent foramen ovale; VSD - Ventricular septal defect.
*The groups were unmatched.

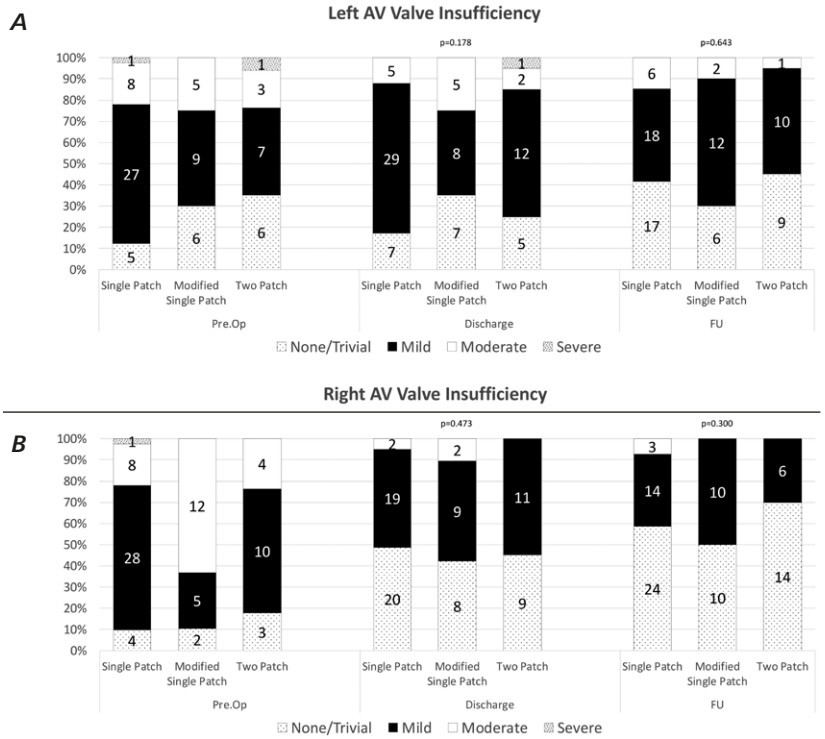

Figure 1

*Bar charts representing progression of left (**A**) and right (**B**) atrioventricular valve insufficiency for each group. The contemplated stages are: status previous to surgery (Pre.Op); status in the last in-hospital evaluation (Discharge); and status at the last follow-up (FU). The numbers inside the bars are the number of patients with the respective level of insufficiency. On top of each stage is the p value for the comparison between groups on the respective stage. The groups were unmatched.*

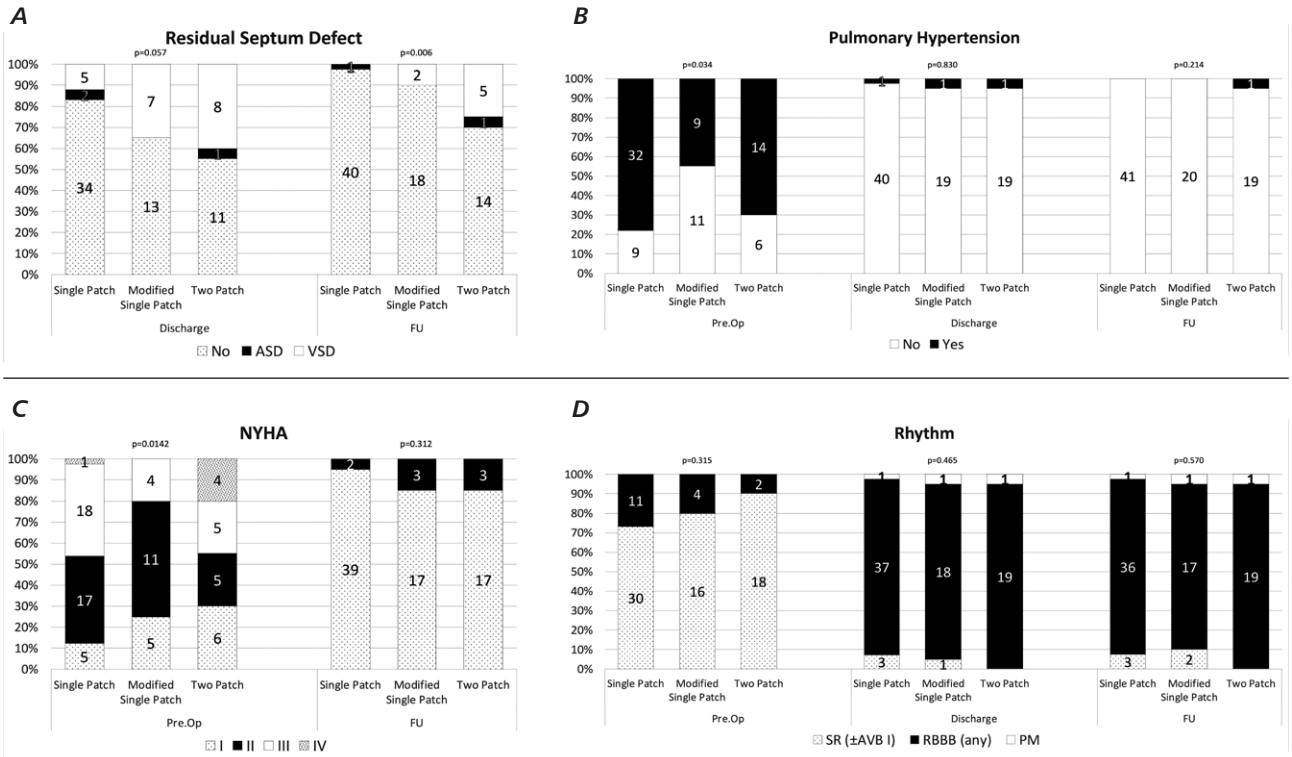

Figure 2

*Bar charts representing progression of residual septal defects (**A**), pulmonary hypertension (**B**), heart rhythm (**C**) and New York Heart Association (NYHA) class. The contemplated stages are: pre-operative status (Pre.Op); status in the last in-hospital evaluation (Discharge); and status at the last follow-up (FU). The numbers inside the bars are the number of patients with the respective level of insufficiency. On top of each stage is the p value for the comparison between groups on the respective stage. The groups were unmatched.*
ASD - Atrial septal defect; AVB I – First-degree atrioventricular block; PM - Permanent pacemaker; SR - Sinus rhythm; RBBB - Right bundle branch block; VSD - Ventricular septal defect.

the SPG (Figure 2A). During follow-up, the incidence, due to spontaneous closure, reduced to 30%, 10% and 2.4% (p=0.006), respectively. None of the residual defects was hemodynamically relevant.

Pulmonary hypertension

Prior to surgery, PHT was present in 68% of the patients, with the incidence varying between groups (see Table 1 for details). After surgery, PHT crisis occurred in 45% of the TPG, 20% of the mSPG and 17% of the SPG. At discharge, 1 patient in each group had PHT (Figure 2B). At follow-up, only 1 patient (from the TPG) remained with high pressure levels.

Statically significant differences between groups were only found in the preoperative frequency of PHT (p=0.034).

Rhythm

In the early postoperative period, temporary, but clinically relevant, arrhythmic episodes (such as heart block and junctional ectopic tachycardia) occurred in 30% of the TPG and SPG; in the mSPG the frequency was 15% (Figure 1D).

Permanent pacemaker (PM) implantation after surgery was 2.4% in the SPG and 5% in both mSPG and TPG.

Right bundle branch block (RBBB) was the most common permanent rhythm anomaly in all groups; overall it was 91%. All the 17 patients that previously to surgery had an AV block (AV block I or RBBB) sustained RBBB after surgery. New RBBB after procedure was 89%. This outcome was kept during follow-up, similarly, in all groups.

No statically significant differences were found between groups, regarding rhythm anomalies.

Survival, functional status and other follow--up data

Mean follow-up time was different between groups: 17.1 ± 15.8 months in TPG, 30.6 ± 34.7 months in mSPG and 57.2 ± 39.7 months in SPG (p<0.001).

No patient died in the perioperative period or during the follow-up.

More than 85% of the patients presented NYHA functional class I at follow-up, without meaningful differences between the groups; the remaining were at class II (Figure 2C).

LVOT obstruction was found in 3 patients: 2 in the SPG and 1 in the TPG. In the 2 patients from the SPG the obstruction needed surgical correction; these were the only AVSD-related reoperations that occurred in all groups during follow-up.

During follow-up, 1 case of endocarditis was found in the SPG, 28 months after surgery, and 1 case of

sepsis occurred in the mSPG, 16 months post-surgery. Four patients in SPG, 4 in the TPG and 1 in the mSPG were hospitalized due to respiratory infection, at least once; all but one, had Down syndrome.

COMMENT

The definitive treatment for complete AVSD is the surgical repair. Several techniques have emerged to resolve this challenging congenital heart defect, with three of them being commonly used: single patch technique, two-patch, and modified single patch technique. None have shown superiority over the others and, regardless of the technique, several post-operative complications (left AV valve dysfunction, conduction system disturbance, postoperative pulmonary hypertensive crisis etc.) are known to occur.

All three techniques are being currently used at our institution. The technique of repair is generally decided intraoperative according to the anatomy and the surgeon's preferences, which outlines the reason for the different population baseline characteristics between the groups. A small VSD usually leads to smaller left to right shunt and, consequently, less PHT and symptoms. This allows the patients to mature more before needing surgery (either PA banding or defect repair), which leads to a faster and less complicated recovery. The modified single patch technique was mainly used when the VSD was small and this very important anatomical advantage was the reason why mSPG presented overall better results. Despite this, in general, all three techniques resulted in satisfactory surgical and clinical results, which improved during follow-up. Some patients needed reoperation in the early postoperative period due to severe left AV valve regurgitation. In particular, the number of intraoperative revisions in the TPG and mSPG was high. This is justified by the small number of patients and the initial learning curve. Although one of the patients was discharged with unsatisfactory results regarding the left AV valve, in the follow up the overall patient population showed spontaneous improvement with no cases of severe left AV valve insufficiency.

Right AV valve regurgitation was greatly reduced with surgery, which was also favored from the change in pressure after closing the intracardiac shunts, positioning the right AV-valve in the low-pressure circulation. Small and hemodynamically not relevant residual septal defects were not uncommon in the early postoperative period, being most frequent in the TPG; this may be related to the additional location for possible residual AVSD that the two-patch technique presents. With time, the residual defects tended to close spontaneously, and the differences found between groups (favoring the single patch technique) may result from the different follow-up times (longer in the SPG). PHT improved, as anticipated, greatly during recovery. Despite high frequencies of PHT before surgery and PHT crisis in the first days after operation, only a minority of patients showed signs of elevated pulmonary pressure at discharge, that persisted in only one patient at follow-up. All structural and physiologic improvements reflected in the functional status, with at least 90% in NYHA class I, and the remaining in class II, and in the survival, with no mortality.

Permanent PM implantation and LVOT obstruction are serious complications associated with AVSD repair. Global PM implantation was 3.7% (with 1 case in each group). Regardless of the technique, RBBB was much more frequent. This conduction anomaly is underreported, since it is considered less problematic than complete AV Block. However, an increased risk of mortality has been found in adults with RBBB (both in the general population and in patients with heart disease) and it can be expected that this may also have impact on the child's long-term prognosis.[9,10] LVOT obstruction also occurred in 3.7% of the patients (2 in the SPG; 1 in the TPG), and 2 of those patients required surgical treatment.

After literature research, we found the incidence of the major complications after surgical correction for complete AVSD in our study (PM implantation, AV-Valve regurgitation, LVOT obstruction, etc) comparable with the ones in early reports.[5,7,8,11]

Infections during the recovery of a surgery is a well-known risk factor for mortality. For this reason, antibiotics are in many cases initiated in presence of suggestive symptoms/signs or elevated inflammatory markers, without a previous clinical diagnosis of infection. Detailed information about infections after complete AVSD repair is not always present in the literature. Reports of sepsis ranging from 5.3 to 7.5% and respiratory infection from 8.7 to 59% were found[12,13,14,15]; these also report a few cases of infection-related mortality. By direct comparison we obtained less cases of sepsis. Long ventilation demand, long ICU and hospital stay on a very young population with 84% Down syndrome patients is probably the reason for the number of infections. Supporting this idea is the fact that both TPG and SPG, with a more fragile population and longer ventilation length and ICU stay, acquired more infections, comparing to the mSPD. On the other hand, these differences may exist due to a low number of patients. All cases were treated and no associated mortality occurred. Concerning infections during follow-up, respiratory infections were the most frequent. Endocarditis/sepsis occurred in 2 patients of the SPG (2.5% globally, 4.9% within group), more than 1 year from surgery. The patents were hospitalized, properly treated and recovered good functional status without sequels.

Many surgical groups have compared the different techniques of repair for CAVSD, with none showing definitive superiority over the other. We added the two patch technique to our routine in order to expand our ability to better treat the variety of surgical anatomies of the AV canal, with the surgeon deciding, intraoperative, which technique would best fit the patient. The mandatory echocardiography in the operating room helped us revise unsatisfactory results immediately, reducing the need for late re-operations and, probably, also the overall length of

hospital stay. Furthermore, residual AV-Valve insufficiency tended to reduce during follow up and small septal defects to close spontaneously, allowing the patients to achieve a good functional status with excellent survival and high freedom from re-intervention over time.

Study limitations

This report shows several limitations like the low number of patients and the relative short follow-up times of the mSPG and the TPG. Furthermore, the groups were not matched.

Disclosures and Freedom of Investigation

The authors report no conflicts of interest regarding the content of this study.

REFERENCES

1. Jacobs JP, Burke RP, Quintessenza JA, Mavroudis C. Congenital Heart Surgery Nomenclature and Database Project: Atrioventricular canal defect. Ann Thorac Surg 2000; 69:S36-S43.
2. Marino B. Congenital heart disease in patients with Down's syndrome: Anatomic and genetic aspects. Biomed Pharmacother 1993; 47:197-200.
3. Karl TR, Provenzano SC, Nunn GR, Anderson RH. The current surgical perspective to repair of atrioventricular septal defect with common atrioventricular junction. Cardiol Young 2010; 20(3):120-7.
4. Rastelli GC, Ongley PA, McGoon DC. Surgical repair of complete atrio-ventricular canal with anterior common leaflet undivided and unattached to ventricular septum. Mayo Clin Proc 1969; 44:335-341.
5. Sabine H, Daebritz, MD. Correction of Complete Atrioventricular Septal Defects With Two Patch Technique. Oper Tech Thorac Cardiovasc Surg 2004; 9:208-220.
6. Geoffrion TR, Singappuli K, Murala JSK. A review of the Nunn modified single patch technique for atrioventricular septal defect repair. Transl Pediatr. 2018; 7(2):91-103.
7. Pan G1, Song L, Zhou X, Zhao J. Complete atrioventricular septal defect: comparison of modified single-patch technique with two-patch technique in infants. J Card Surg 2014; 29(2):251-5.
8. Li D, Fan Q, Iwase T, Hirata Y, An Q. Modified Single-Patch Technique Versus Two-Patch Technique for the Repair of Complete Atrioventricular Septal Defect: A Meta-Analysis. Pediatr Cardiol 2017; 38(7):1456-1464.
9. Xiong Y, Wang L, Liu W, Hankey GJ, Xu B, Wang S. The Prognostic Significance of Right Bundle Branch Block: A Meta-analysis of Prospective Cohort Studies. Clin Cardiol 2015; 38(10):604-13.
10. Harkness WT, Hicks M. Rhythm, Bundle Branch Block, Right (RBBB). StatPearls [Internet] 2018.
11. Harmandar B1, Aydemir NA, Karaci AR, Sasmazel A, Saritas T, Bilal MS, Yekeler I. Results for surgical correction of complete atrioventricular septal defect: associations with age, surgical era, and technique. J Card Surg. 2012 Nov;27(6):745-53.
12. Deraz S, Ismail M. Single patch technique versus double patch technique in repair of complete atrioventricular septal defect. The Egyptian Heart Journal 2013. 66(2): 177-182.
13. Sivalingam S, Krishnasamy S, Al-Fahmi NA et al. Early and midterm outcome of complete atrioventricular septal defect (AVSD) in a single institution. Indian J Thorac Cardiovasc Surg 2013. 29:223–229.
14. Atz AM, Hawkins JA, Lu M et al. Surgical management of complete atrioventricular septal defect: associations with surgical technique, age, and trisomy 21. J Thorac Cardiovasc Surg 2011. 141:1371–1379.
15. Pan G, Song L, Zhou X et al. Complete atrioventricular septal defect: comparison of modified single-patch technique with two-patch technique in infants. J Card Surg 2014. 29:251–255.

ARTIGO ORIGINAL ORIGINAL ARTICLE

CONCOMITANT ASCENDING AORTIC REPLACEMENT AND AORTIC VALVE REPLACEMENT USING RAPID DEPLOYMENT BIOPROSTHESIS

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Abstract

Introduction: The association between aortic valve disease and dilatation of the ascending aorta is well known and concomitant surgery is recommended when the aortic diameter is higher than 45mm. The use of the rapid deployment valves allows less cross-clamping and cardiopulmonary bypass times for both isolated and combined procedures in comparison to regular valves.

We describe our initial experience of concomitant aortic valve and the ascending aortic replacement, using the rapid deployment valve Edward Intuity EliteTM.

Case presentation: All patients were male, with a mean age of 72-years-old. The mean cross-clamping time was 48 minutes, with a mean cardiopulmonary time of 61 minutes. The mean time of ICU stay was 4 days. All the patients had follow-up 1 and 3 months after discharge and were doing well.

Conclusions: The rapid deployment aortic valves have recognized advantages in aortic valve replacement. Our small experience reinforces that replacement the ascending aortic and aortic valve with this prothesis is one procedure that can benefits from generalization without increased risks and with potentially better clinical outcomes. Larger cohort studies would allow clarification over this subject.

INTRODUCTION

The association between aortic valve disease and dilatation of the ascending aorta is well known and concomitant ascending aortic replacement is recommended at the time of aortic valve replacement surgery if the diameter is greater than 45mm.[1,2,3]

Among the numerous advantages associated to the use of the rapid deployment valves, cross-clamp time (CXT) and cardiopulmonary bypass time (CBPT) reductions, for both isolated and combined procedures is of paramount importance.[4]

The ability to use this device in complex composite grafts or separated grafts for addressing concomitant ascending aortic pathology seems particularly helpful and allows important time reductions in comparison to regular valves.[5,6,7]

We describe our initial experience of aortic valve replacement and ascending aortic replacement, using the rapid deployment valve Edward Intuity Elite™.

CLINICAL CASES

Since the beginning of our rapid deployment bioprosthesis program, three patients were submitted to separated aortic valve replacement and ascending aortic replacement, using the valve Edward Intuity Elite™ and Dacron tube, in the same surgical procedure (Table 1 and 2).

CASE 1

A 64 years-old male, with hypertension as sole comorbidity, presented with dyspnoea of progressive worsening with efforts. Coronary angiography was normal. Echocardiogram showed: normal left ventricular function (LVF); aortic orifice area of 0.9cm^2; peak gradient of 85mmHg and medium gradient of 54mmHg; without any other valvular abnormalities. Thoracic computed tomography (CT) scan revealed the following aortic dimensions: root 34mm; ascending 47mm; arch 43mm.

Table 1 **Preoperative demographics for patients undergoing aortic valve replacement with Intuity prothesis and aorta replacement.**

Patient	Age	BMI	EF	Euroscore II	Aortic valve Patology	Aneurism (mm)
1	64	29,4	>50%	1,84	Stenosis	47
2	80	32,7	35%	3,90	Stenosis	53
3	72	30	>50%	2.30	Stenosis	48

BMI - Body mass index; EF - Ejection Fraction;

Table 2 **Intraoperative demographics for patients undergoing aortic valve replacement with Intuity prothesis and aorta replacement.**

Patient	1	2	3
Aortic prosthesis/tubular	25/ 26	27/ 30	23/ 28
CPBT[1]	51	69	63
CXT[1]	39	54	52
UCI[2]	2	7	3
Aminergic support	No	Yes	No
Drainage 24h[3]	300	1100	100
Hospital stay[2]	6	10	6
Complications	AF	Hemostasis review; AF	No
Follow-up	OK	OK	OK

1 – Min; 2 – Days; 3 – mL; CPBT - Cardiopulmonary bypass; CXT – cross-clamp time; AF – Atrial fibrillation.

The patient was submitted to an aortic valve and ascending aorta replacement, by median sternotomy. Under cardiopulmonary bypass, the distal ascending aorta was cross clamped. After aortotomy, the dilated aortic segment and the calcified and stenotic tricuspid aortic valve were removed. A biological rapid deployment aortic valve Edward Intuity Elite™ 25 was implanted followed by a Dacron tubular prosthesis number 26.

The CXT was 39 min with a CBPT of 51 min. Intraoperative transesophageal echocardiogram demonstrated normofunctional prosthesis, no paravalvular leak and a mean gradient of 5 mmHg.

In the post-operative the patient had one episode of atrial fibrillation (AF), converted to sinus rhythm with medical therapy, without other events.

CASE 2

80-years-old male, with hypertension, persistent AF and COPD, with recurrent hospitalizations for decompensated heart failure. Echocardiogram showed severe aortic stenosis, with moderate LVF (35%). The CT revelled an ascending aortic aneurysm with 53mm, with calcification but able for clamping. The coronarography was normal.

He was submitted to aortic valve replacement and replacement of the ascending aorta using the same method described in the Case 1, but instead we used an aortic valve Edward Intuity Elite™ 27 and a tubular prosthesis 30. The CXT time was 54 min with a total CBPT of 66 min. Intraoperative transoesophageal echocardiogram demonstrated well positioned prosthesis, no paravalvular leak and a mean gradient of 3 mmHg.

The patient had a significant loss of blood in the immediate post-operative, and went back to the operation room for haemostasis due to bleeding. After, the patient had an uneventful recovery.

CASE 3

A 72-years-old male, with diabetes and high blood pressure, presented with 2 recent syncopal episodes. Coronary angiography showed a lesion of 50% in the circumflex artery, without other lesions. Echocardiogram showed a normal biventricular function and an aortic valve with severe aortic stenosis with moderate insufficiency. CT scan showed a tubular thread ascending with 48mm.

The patient was submitted to an aortic valve and ascending aorta replacement, using a biological rapid deployment aortic valve, Edward Intuity Elite™ 23 and a Dacron tube 28.

The CXT was 52 min with a CBPT of 63 min. Intraoperative transesophageal echocardiogram demonstrated preserved biventricular function and a well-positioned prosthesis without leaks.

The patient had an uneventful recovery.

All the patients had *follow-up* at 1 and 3 months after discharge and were doing well.

DISCUSSION AND CONCLUSION

The development of rapid deployment aortic valves proved to be a great advancement for aortic valve surgery. Reduced CXT and CBPT for both isolated or combined procedures, superior hemodynamic performance, facilitator of broader minimally invasive procedures and reduced aortic manipulation are some of the recognized advantages of this device.[3,4]

If these advantages are well established should we not explore them to their full potential?

Although with some described drawbacks such as, paravalvular leaks and higher pacemaker implantation rates, from a conceptual point of view, separated ascending aortic replacement and aortic valve replacement using Edward Intuity Elite™ is one of the procedures that can benefit from generalization without increased risks. In fact, such procedures or even other more complex evolving the aortic root have already been successful performed before.[6,7]

Our small experience reinforces that option as a fast and safe way to simplify these complex procedures with potentially better clinical outcomes. The mean CXT and CBPT were 48 and 61 minutes, respectively and the bioprosthesis hemodynamic performance was excellent. There were no paravalvular leaks or permanent conduction disorders leading to pacemaker implantation. Nevertheless, these operative results did not translate in reduced ICU stay. At 1 and 3-months *follow-up* after hospital discharge all the patients were doing well, with echocardiogram without abnormalities and with normal valve function.

In conclusion, aortic valve and ascending aortic replacement, using a rapid deployment bioprosthesis might be a safe and useful approach.

Our study has several limitations, including the small sample size, short follow-up, and retrospective nature. Future investigations with more patients and a longer follow-up will be required to confirm the outcomes of this approach.

REFERENCES

1. Lee, S. H., Kim, J. B., Kim, D. H., Jung, S. H., Choo, S. J., Chung, C. H., & Lee, J. W. (2013). Management of dilated ascending aorta during aortic valve replacement: Valve replacement alone versus aorta wrapping versus aorta replacement. Journal of Thoracic and Cardiovascular Surgery, 146(4), 802–809.
2. Borger, M. A., Preston, M., Ivanov, J., Fedak, P. W. M., Davierwala, P., Armstrong, S., & David, T. E. (2004). Should the ascending aorta be replaced more frequently in patients with bicuspid aortic valve disease? Journal of Thoracic and Cardiovascular Surgery, 128(5), 677–683.
3. Erbel R., Aboyans V., Boileau C., Bossone E., Bartolomeo R., Eggebrecht H., … Vrints C. (2014). 2014 ESC Guidelines on the diagnosis and treatment of aortic diseases. European Heart Journal (2014) 35, 2903.
4. Gersak, B., Fischlein, T., Folliguet, T. A., Meuris, B., Teoh, K. H. T., Moten, S. C., … Glauber, M. (2016). Sutureless, rapid deployment valves and stented bioprosthesis in aortic valve replacement: Recommendations of an international expert consensus panel. European Journal of Cardio-Thoracic Surgery, 49(3), 709–718.
5. Silva, I. A., Corso, R. B., Santos, M. V. N., Souza, H. J. B. de, & Pina, G. K. S. (2019). First Two Brazilian Cases: Correction of Ascending Aortic Aneurysm and Aortic Valve Stenosis with Sutureless/Rapid Deployment Aortic Prosthesis. Brazilian Journal of Cardiovascular Surgery, 34(3), 2018–2019.
6. Lio, A., Ferrarini, M., Miceli, A., & Glauber, M. (2018). Sutureless Prosthesis Implantation and Ascending Aorta Replacement Through a Ministernotomy Approach. Innovations: Technology and Techniques in Cardiothoracic and Vascular Surgery, 13(3), 239–241.
7. Gennari, M., Agrifoglio, M., & Polvani, G. (2017). Composite graft using an Edwards Intuity Elite rapid deployment bioprosthesis for aortic root replacement. Journal of Cardiac Surgery, 32(3), 193–195.

ARTIGO ORIGINAL ORIGINAL ARTICLE

CARDIAC SURGERY IN PATIENTS WITH DIALYSIS-DEPENDENT END STAGE RENAL FAILURE: SINGLE CENTRE EXPERIENCE

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Abstract

Background: Patients under dialysis have a high cardiovascular risk and they are at increased risk when submitted to cardiac surgery.

Aim of the study: to evaluate morbidity, early and late mortality, and predictive factors of mortality in patients under dialysis who underwent cardiac surgery.

Methods: A retrospective observational study was performed including all dialysis dependent patients who underwent cardiac surgery (coronary, valvular or combined procedures) in our institution between 2007 and 2014. A population of 95 consecutive patients was obtained (no exclusions). Perioperative variables and predictors of mortality were analysed and the endpoints were early and late mortality. Propensity score matching, with a control group of patients with creatinine clearance >90mL/min, was performed by logistic regression, with a 1:1 matching. Kaplan Meier curves were performed for late mortality.

Results: Early mortality was 9.4% (EuroSCORE II 4.1%). In univariate analysis, mean time of cardiopulmonary bypass (CPB) ($p=0.016$) and EuroSCORE II ($p=0.02$) were related with early mortality. In a multivariate analysis model, combined procedures (OR 138.09; CI95% 1.82-10498.4; $p=0.03$) and CCS (Canadian Cardiovascular Society) 3-4 (OR 70.951; CI 95% 1.32-3810.11; $p=0.037$) were predictors of mortality. In multivariable analysis, CPB time >152 min was a predictor of early mortality ($p=0.001$). After propensity score matching, 30 day, one year and late mortality were higher in the dialysis group.

Conclusions: Early and late mortality were significantly higher in dialysis dependent patients. Predictive factors of mortality were CPB time and EuroSCORE II in univariable analysis, and CCS 3-4 and combined procedures in multivariable analysis.

INTRODUCTION

The incidence and prevalence of patients with renal failure who need kidney replacement therapy is increasing. In Portugal there were 11738 patients (1135 patients per million population) undergoing hemodialysis in 2016.[1] Dialysis dependent patients develop coronary and valvular disease earlier and more frequently than the general population. In 2016, dialysis patients global mortality was 13.09% and death due to cardiovascular disease was 24.6%.[1]

Accounting for this cardiovascular burden, non-specific and specific risk factors are identified, including vascular and valvular calcifications and left ventricular hypertrophy. Consequently, surgical risk is greater.[2] However, studies have been conflicting regarding mortality and its predictors in dialysis patients. Thus, it continues to be unclear which patients benefit from surgery and when is surgical risk prohibitive.

The purpose of this study was to characterize end stage renal disease (ESRD) patients under dialysis who underwent cardiac surgery in one institution, to evaluate morbidity, early and late mortality, time to discharge and determine predictive factors for these outcomes.

MATERIALS AND METHODS

A retrospective observational study was performed including all ESRD-dialysis dependent patients who

underwent cardiac surgery in one institution between 2007 and 2014. No exclusion factor was applied. The population was characterized regarding perioperative variables, including EuroSCORE I and II and the endpoints were early (≤30days after surgery) and late mortality (mortality during follow up). Mortality was obtained from consultation of a national register, with a follow up rate of 100%. Mean follow-up was 3.6 ±2 years.

Patients

A population of 95 ESRD-dialysis dependent patients was obtained (96% haemodialysis; 4% peritoneal dialysis). Mean age was 65±11 years (range 33 to 88 years), 81% were male. Preoperatively, 33% had previous history of acute myocardial infarction; 35% had an ejection fraction <50%. (Table 1)

Table 1 **Cardiovascular risk factors and other preoperative characteristics. Extracardiac vascular disease: Cerebrovascular disease, peripheral vascular disease, claudication.**
(CCS: Canadian Cardiovascular Society; NHYA: New York Heart Association)

Male gender	77	81%
Diabetes mellitus	35	36.8
Dyslipidaemia	43	45.3
Smoking	25	26.3
Overweight/obesity	48	50.5
Hypertension	79	83.2
Extracardiac vascular disease	32	33.7
Cerebrovascular disease	18	18.9
Poor ejection fraction (<30%)	3	3.2
NYHA III/IV	23	24.2
CCS 3/4	22	23.2
Urgent/emergent operation	17	18%

Patient management protocol

Dialysis patients are always referred to the Nephrology service on admission so that a pre-operative dialysis session can be performed. Post-operative surveillance is maintained by the nephrology team. This ensures optimal hydroelectrolytic and metabolic status at the time of surgery and adequate management in the perioperative period.

Statistical analysis

Statistical analysis, including univariable and multivariable analysis (logistic regression) to identify predictors of early mortality were performed using SPSS v. 20. A p value of 0.05 or less was considered significant. A propensity score matching model was performed by logistic regression, including all variables statistically and clinically relevant (age, sex, comorbidities, preoperative status, type and characteristics of surgical procedure and mortality) with a 1:1 matching. The control group included all patients submitted to cardiac surgery during the same time frame with estimated creatinine clearance >90mL/min based on Cockcroft Gault equation (n=2144). Kaplan Meier curves were performed for late mortality.

RESULTS

Regarding timing of surgery, 77 were elective and 17 were urgent/emergent. Reoperations accounted for 8.5% of total of surgeries. Procedures performed are presented in Table 2.

Table 2 **Procedures performed**

Main procedures	N =95	%
Coronary artery bypass graft	42	44%
Valvular surgery Isolated aortic valve Multiple valve	33 22 11	35% 23% 12%
Combined valvular and coronary surgery	11	12%
Other procedures	9	9%

Cardiopulmonary bypass (CPB) was used in 63.2% of surgeries. Mean time of CPB was 110 ± 50 minutes (minimum 20 minutes, maximum 256 minutes).

Revascularization included 2.3±0.96 grafts/patient, arterial grafts in 92.7% and complete revascularization in 74.1%. Among coronary artery bypass graft (CABG) surgeries, 71.4% (30 cases) were performed off pump.

Valvular surgery involved 48 prosthetic valves (44% of which were mechanical) and 11 valve repairs. Two aortic valve replacements were performed off pump (transapical approach).

Early mortality was 9.4% (EuroSCORE 9.5%, EuroSCORE II 4.1%) (Table 3). The median time of in-hospital stay after surgery was 8 days. One-year mortality was 18.9%. Mortality at the end of follow-up (3.6 ±2 years) was 33.7%.

Table 3 **EuroSCORE I and II and early mortality.**
(CABG: coronary artery bypass graft)

	CABG (n=42)	Valvular surgery (n=33)	All patients (n=95)
EuroSCORE I (mean)	7.5%	10.7%	9.5%
EuroSCORE II (mean)	3.4%	5.3%	4.1%
Early mortality (%)	4.8%	9.1%	9.4%

Regarding CABG, early mortality was 0% (0 out of 30) in off pump surgery and 14% when on pump (1 death out of 7 patients). In univariate analysis, mean time of CPB ($p=0.016$) and EuroSCORE II ($p=0.02$) were related with early mortality (Table 4).

Table 4 **Univariable analysis of early mortality**
(CCS: Canadian Cardiovascular Society; CPB: cardiopulmonary bypass; NHYA: New York Heart Association)

	p
Diabetes mellitus	0.282
Dyslipidaemia	0.694
Hypertension	1.000
Extracardiac arteriopathy	0.713
Low ejection fraction (<30%)	1.000
NYHA III/IV	0.035
CCS 3/4	0.428
Mean time of CPB	0.016
EuroSCORE II	0.02

In a multivariate analysis model, combined procedures and CCS 3-4 were predictors of mortality with an OR 138.09 (CI95% 1.82-10498.4; $p=0.03$) and OR 70.951 (CI 95% 1.32-3810.11; $p=0.037$) respectively (Table 5).

With ROC curves analysis, CPB time >152 minutes was the cut-off value for increased risk of early mortality. In multivariable analysis, CPB time >152 min was a predictor of early mortality ($p=0.001$).

Propensity score matching

In order to compare mortality in this population with a control group with normal renal function (estimated creatinine clearance > 90ml/min), propensity score matching 1:1 was performed. The characteristics of the control (normal renal function) group are presented in table 6.

Table 6 **Normal renal function (control) group.**
(CCS: Canadian Cardiovascular Society; CPB: cardiopulmonary bypass; NHYA: New York Heart Association)

Male gender	77	81%
Diabetes mellitus	44	46%
Smoking	11	12%
Overweight/obesity	52	55%
Hypertension	64	67%
Extracardiac arteriopathy	10	11%
Cerebrovascular disease	17	18%
Poor ejection fraction (<30%)	2	2%
NYHA III/IV	23	24%
CCS 3/4	16	17%
Urgent/emergent operation	14	15%
Age (mean)	62 years	
EuroSCORE II (mean)	2.1%	
CBP time (mean)	95 minutes	

After propensity score matching, 30 day and 1 year mortality were significantly higher in the dialysis group comparing with the control group. (Table 7) Analysis of

Table 5 **Multivariable analysis of early mortality.**
(BMI: body mass index; CI: confidence interval; CCS: Canadian Cardiovascular Society; CPB: cardiopulmonary bypass; NHYA: New York Heart Association; OR: odds ratio)

Variables	OR	95% CI	*p*
CPB	1.586	0.07-34.32	0.77
Urgent/emergent	1.135	0.14-9.35	0.91
Combined procedures	138.090	1.82-10498.4	0.03
CCS 3-4	70.951	1.32-3810.11	0.04
Diabetes mellitus	1.757	0.23-13.68	0.59
Extracardiac arteriopathy	0.164	0.01-2.63	0.20
NHYA III/IV	5.817	0.61-55.77	0.13
Ejection fraction	0.000	0-0	1.00
BMI	1.166	0.94-1.45	0.16
Age	1.028	0.92-1.14	0.61
Female	0.221	0.01-3.8	0.30
Cerebrovascular disease	0.806	0.01-43.97	0.92
Pulmonary disease	2.177	0.1-49.19	0.62
Coronary disease	1.392	0.04-43.47	0.85

Table 7 **Comparison of early and one year mortality in the dialysis *vs.* the control (normal renal function) group.**

	Dialysis group	Control group	*p*
Early mortality (≤30days)	9.4%	0%	<0.001
1-year mortality	18.9%	3.2%	<0.001

late mortality using Kaplan Meier curves also demonstrated a higher mortality of the dialysis group (figure 1).

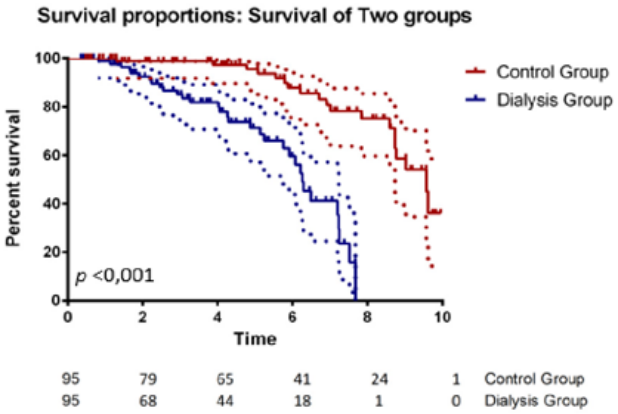

Figure 1 *Kaplan Meier curves for survival of control (normal renal function) and dialysis groups after propensity score matching (time in years).*

DISCUSSION

Overall mortality due to cardiovascular disease is more frequent in dialysis-dependent patients (30% to 50%) than in an age-corrected control population (less than 15%).[3,4]

Coronary heart disease affects 30%–60% of patients with ESRD.[5] This high incidence is related with predisposing factors such as hyperlipidaemia, hypertension, anaemia, fluid overload, platelet dysfunction and disturbances of calcium phosphate metabolism.[6] ESRD patients usually present multivessel disease, and present with proximal lesions, extensive calcifications, or diffuse disease.[5]

Due to the coexisting noncardiac morbidities, the deleterious consequences of renal disease and dialysis, perioperative results and long term mortality of cardiac surgery have been worse in dialysis patients.[3] Furthermore, indications and referral for operation may often be delayed in patients with ESRD and this may also contribute to the high perioperative mortality.[7] However, although perioperative risk of dialysis-dependent patients is higher, it is not prohibitive, and good candidates for surgery can be identified.[8]

According to Ko *et al.*, the overall operative mortality of dialysis patients reported in the literature was 9% in 1993. In isolated CABG procedures it was 9%, 12% in valvular operations and 13% in combined CABG with valvular operations.[9]

Horst *et al.* reviewed available literature in 2000 obtaining 863 patients over 30 years and described a perioperative mortality rate for isolated CABG, isolated cardiac valve operation, and combined procedures of 8.9%, 19.3%, and 39.5%, respectively. Overall perioperative mortality rate was 12.5% combining all cardiac surgical procedures with CPB.[10]

More recently, Nicolini et. al. revised 18 reports with a total of 1725 dialysis patients submitted to heart surgery and presented a mean perioperative mortality rate of 13.3% (range 0 to 36.7%).[11]

Takami *et al.* reported an in-hospital mortality of 9.8% in a group of 245 haemodialysis dependent patients.[12] In a study with 45 patients, thirty-day mortality was 13.3% and late mortality was 46.6%.[13]

Yamauchi *et al.* compared 1,300 HD-dependent chronic renal failure patients with 18,387 non-HD patients submitted to isolated CABG and described a 30-day mortality of 4.8% *vs.* 1.4% in the HD and non-HD groups, respectively.[14] Operative mortality and major complications were also more frequent in the HD group (23.1% *vs.* 13.7%).[14]

In a study including 5308 patients who underwent valve surgery, including 224 dialysis dependent patients, the in-hospital mortality rate for the entire cohort was 5.7% (n = 304) and it was significantly higher for those under dialysis before surgery (18.3% *vs* 5.2%; $p < .0001$).[15]

Deutsch *et al.* obtained a 30-day mortality of 17.6% in a group of 204 patients. The highest mortality rates occurred in patients undergoing combined procedures.[2]

Rahmanian *et al.* in a series of 245 patients with end-stage kidney failure requiring dialysis stratified mortality by procedure and obtained the highest mortality rate in patients undergoing single/multiple valve procedures (17.1%), followed by combined valve/CABG (12.8%), isolated CABG (10.3%), and aortic (9.1%) procedures. Overall hospital mortality was 12.7% and the EuroSCORE was 18%.[16]

Yamamura studied 76 dialysis patients and reported an overall in-hospital mortality rate of 17.1%. In patients undergoing CABG the hospital mortality rate was 13.8%, in aortic valve replacements (AVR) it was 12.5% and in AVR plus CABG 33.3%.[17]

Therefore early mortality in our centre (9.4%) is comparable to that reported in literature and to EuroSCORE. Mortality according to procedure (4.8% for isolated CABG and 9.1% for isolated valvular surgery) is favourable compared to other reports in literature.

Rahmanian *et al.* also described 1-year, 3-year, and 5-year survival of 72.3%± 3.3%, 53.3%± 4.0%, and 39.0% ± 4.5% respectively in a series of 214 discharged patients.[16]

Yamamura obtained a 5-year overall survival rate of 39% ±8%.[17] The 1-year survival rate for isolated CABG

was 77.0% ± 0.7 %; for isolated AVR was 79.0% ±0.8% and for concomitant surgery was 21% ±18%. In this last group, survival was significantly poorer.[17]

One year (18.9%) and late mortality in our series (21.9%) is in the range of that reported by other authors.

Regarding predictors of mortality, many have been pointed out in different studies. In the study by Yamauchi *et al.*, age, chronic obstructive pulmonary disease, peripheral artery disease, congestive heart failure, arrhythmia, preoperative inotropic agent requirement, New York Heart Association class IV, urgent or emergent operation, poor left ventricular function, aortic valve regurgitation (>2), and mitral valve regurgitation (>3) were indicated as preoperative predictors of operative mortality in the dialysis group.[14]

Horst et. al. described duration of dialysis equal to or longer than 60 months, and NYHA class IV as being associated with substantially increased relative risk for perioperative death.[10]

Yamamura *et al.* showed that age higher than 70 years, low-output and concomitant surgery were significant risk factors for mortality in a univariate logistic analysis. The multivariate logistic analysis described concomitant surgery (odds ratio 4.37, $p<0.007$) as the only significant risk factor for mortality.[17] Horst *et al.* also reported combined procedures as a risk factor for mortality based on a univariate logistic analysis.[10] Rahmanian *et al.* identified peripheral vascular disease as an independent predictor for mortality.[16]

NYHA class IV and emergent operation were appointed as risk factors for mortality by Ko *et al.* based on a univariate logistic analysis.[9] Takami *et al.* identified only diabetes mellitus as an independent predictor of hospital mortality with an odds ratio of 2.74.[12]

Age, blood product usage and postoperative pulmonary complications have also been described as significant predictors of 30 day mortality and late death.[13]

In the current study, only CCS 3-4 and combined procedures were statistically significant predictors of mortality.

Regarding higher risk compared to general population, Rahmanian *et al.* adjusted for potential confounding factors, and concluded that end-stage kidney failure requiring dialysis was a predictor of hospital mortality (odds ratio, 3.1; $p<0.001$).[16] Propensity score matching found in literature was only used to compare CABG vs PCI or included preoperative non-dialytic renal disease.[18]

After matching our study and control groups, we concluded that 30 day, one year and late mortality were significantly higher in the dialysis group, which did not come as an unexpected result.

Limitations to this study are related to its retrospective and single-centre study and the relatively small number of patients.

CONCLUSIONS

Patients submitted to cardiac surgery who are under dialysis are known to be at higher risk of morbidity and mortality. In our study, early and late mortality was significantly higher in dialysis dependent patients comparing with patients with normal renal function. CPB time and EuroSCORE II were predictive factors of mortality in univariable analysis, whereas CCS 3-4 and combined procedures were related with higher mortality in multivariable analysis. However, EuroSCORE II may underestimate the true risk of this group of patients. This study has limitations due to its retrospective, single-centre and small-sampled methods. A study with a larger number of patients, multicentric and prospective would be the ideal methodology to analyse this population and optimize care according to their specific characteristics.

REFERENCES

1. Macário F, Sociedade Portuguesa Nefrologia: Tratamento substitutivo renal da Doença Renal Crónica Estadio V em Portugal - Relatório Gabinete de Registo da SPN. In.; 2016.
2. Deutsch O, Spiliopoulos K, Kiask T, Katsari E, Rippinger N, Eichinger W, Gansera B: Cardiac surgery in dialysis-dependent patients: impact of gender on early outcome in single-center experience with 204 consecutive cases. Thorac Cardiovasc Surg 2013, 61(1):22-28.
3. Frenken M, Krian A: Cardiovascular operations in patients with dialysis-dependent renal failure. Ann Thorac Surg 1999, 68(3):887-893.
4. Collins AJ: Cardiovascular Mortality in End-Stage Renal Disease. The American Journal of the Medical Sciences 2003, 325(4):163-167.
5. Chang TI, Shilane D, Kazi DS, Montez-Rath ME, Hlatky MA, Winkelmayer WC: Multivessel coronary artery bypass grafting versus percutaneous coronary intervention in ESRD. J Am Soc Nephrol 2012, 23(12):2042-2049.
6. Koyanagi T, Nishida H, Kitamura M, Endo M, Koyanagi H, Kawaguchi M, Magosaki N, Sumiyoshi T, Hosoda S: Comparison of clinical outcomes of coronary artery bypass grafting and percutaneous transluminal coronary angioplasty in renal dialysis patients. Ann Thorac Surg 1996, 61(6):1793-1796.
7. Gharsallah H, Trabelsi W, Hajjej Z, Nasri M, Lebbi A, Jebali M, Ferjani M: Cardiac surgery in patients on hemodialysis: Eight years experience of the Tunisian military hospital, vol. 21; 2010.
8. Bechtel JF, Detter C, Fischlein T, Krabatsch T, Osswald BR, Riess FC, Scholz F, Schonburg M, Stamm C, Sievers HH et al: Cardiac surgery in patients on dialysis: decreased 30-day mortality, unchanged overall survival. Ann Thorac Surg 2008, 85(1):147-153.
9. Ko W, Kreiger KH, Isom OW: Cardiopulmonary bypass procedures in dialysis patients. The Annals of Thoracic Surgery 1993, 55(3):677-684.
10. Horst M, Mehlhorn U, Hoerstrup SP, Suedkamp M, Rainer de Vivie E: Cardiac surgery in patients with end-stage renal disease: 10-year experience. The Annals of Thoracic Surgery 2000, 69(1):96-101.
11. Nicolini F, Fragnito C, Molardi A, Agostinelli A, Campodonico R, Spaggiari I, Beghi C, Gherli T: Heart surgery in patients on chronic dialysis: is there still room for improvement in early

and long-term outcome? Heart and Vessels 2011, 26(1):46-54.
12. Takami Y, Tajima K, Okada N, Fujii K, Sakai Y, Hibino M, Munakata H: Simplified management of hemodialysis--dependent patients undergoing cardiac surgery. Ann Thorac Surg 2009, 88(5):1515-1519.
13. Jayasekera H, Pinto N, Mundy J, Wood A, Beller E, Griffin R, Peters P, Shah P: Cardiac surgery in the presence of dialysis: Effect on mid-term outcomes and quality of life. Heart, Lung and Circulation 2011, 20(2):105-110.
14. Yamauchi T, Miyata H, Sakaguchi T, Miyagawa S, Yoshikawa Y, Takeda K, Motomura N, Tsukihara H, Sawa Y: Coronary Artery Bypass Grafting in Hemodialysis-Dependent Patients: Analysis of Japan Adult Cardiovascular Surgery Database. Circulation Journal 2012, 76(5):1115-1120.
15. Thourani VH, Sarin EL, Kilgo PD, Lattouf OM, Puskas JD, Chen EP, Guyton RA: Short- and long-term outcomes in patients undergoing valve surgery with end-stage renal failure receiving chronic hemodialysis. The Journal of Thoracic and Cardiovascular Surgery 2012, 144(1):117-123.
16. Rahmanian PB, Adams DH, Castillo JG, Vassalotti J, Filsoufi F: Early and late outcome of cardiac surgery in dialysis-dependent patients: Single-center experience with 245 consecutive patients. The Journal of Thoracic and Cardiovascular Surgery 2008, 135(4):915-922.
17. Yamamura M, Mitsuno M, Tanaka H, Kobayashi Y, Ryomoto M, Nishi H, Fukui S, Tsujiya N, Kajiyama T, Miyamoto Y: Risk factors for open heart surgery in hemodialysis patients. Gen Thorac Cardiovasc Surg 2009, 57(5):235-238.
18. Lin CY, Tsai FC, Chen YC, Lee HA, Chen SW, Liu KS, Lin PJ: Correlation of Preoperative Renal Insufficiency With Mortality and Morbidity After Aortic Valve Replacement: A Propensity Score Matching Analysis. Medicine (Baltimore) 2016, 95(9):e2576.

ARTIGO ORIGINAL ORIGINAL ARTICLE

BENEFITS OF CONTINUOUS MONITORING OF PCO_2 OBTAINED FROM A SYSTEM APPLIED TO MEMBRANE OXYGENATOR EXHAUSTION OF THE CARDIOPULMONARY BYPASS CIRCUIT

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Abstract

Objective: To observe the impact of the use of capnography system adapted to cardiopulmonary bypass (CPB). To measure the concordance between values obtained from continuous monitoring of partial pressure of carbon dioxide in membrane oxygenator exhaustion (PeCO2) and the results observed on arterial blood gas test.

Methods: Participated in this study 40 patients submitted to elective cardiovascular surgery with CPB. They were divided into two groups: Group 1, with 20 patients submitted to the surgical procedure using blood gas analysis at intermittent intervals (20 - 30 minutes); Group 2, with 20 patients operated with a capnography system adapted applied to membrane oxygenator exhaustion and blood gas test. A test was used to compare *arterial partial pressure* of carbon dioxide ($PaCO_2$) from group 1 and group 2. In group 2, the strength of the correlation between $PeCO_2$ and $PaCO_2$ was evaluated by a linear regression test. The Bland-Altman method was used to determine the degree of agreement between the two variables.

Results: Average and standard deviation of Group 1's PaCO2 (34.6 ± 7.44) and Group 2's $PaCO_2$ / $PeCO_2$ (36.5 ± 4.42) / (39.9 ± 3.98). There was *no statistically significant difference* in PaCO2 *between* the *groups* ($P = 0.21$). In group 2, $PeCO_2$ and PaCO2 analyzed corrected for esophageal temperature obtained a positive linear correlation ($r = 0.79$, $P < 0.001$), the degree of agreement presented an average 3.47 ± 2.70 mmHg.

Conclusion: The continuous $PeCO_2$ monitoring from cardiopulmonary bypass circuit has a positive impact on the result of PaCO2. This instrument confirms and maintains the carbon dioxide (CO_2) values into reference parameters.

INTRODUCTION

Respiratory acidosis on the cardiopulmonary bypass (CPB) is most related to the retention of carbon dioxide (CO_2) in the membrane oxygenator. Acidosis promotes the appearance of cardiac arrhythmias, decreases inotropic action and causes an increase in pulmonary vascular resistance. Respiratory alkalosis is related to the fast elimination of CO_2 by the oxygenator. This disturbance is crucial for the appearance of neurological complications in the postoperative period of cardiac surgery.[1,2]

The capnography system is a very important method for the monitoring of the patient's respiratory function during surgery. This instrument enables the real-time monitoring of the partial pressure of carbon dioxide (PCO_2) on the expired mixture.[3] The capnographer applied to membrane oxygenator exhaustion of the CPB circuit comprises a system that makes a real-time analysis of the measurement of carbon dioxide elimination in the intervals between arterial blood gases collections. It's presumed that this system assist in the observation and management critical incidents during the cardiopulmonary bypass.[4]

Continuous inline blood gas monitors are not accessible to all cardiac surgery services. An alternative is the adaptation of the capnography to the CPB circuit which permit excellent monitoring of PCO_2, doing it a useful practice and efficient in cardiac surgery.

METHODS

This is a prospective and observational study performed in surgery department of *Instituto Dante Pazzanese de Cardiologia* (IDPC) in São Paulo, SP, Brazil, in June

and July, 2018. The study was approved for the Ethics and Research Committee of the IDPC with CAAE number: 91256018.9.0000.5462.

Participated in this study 40 patients undergoing to elective cardiovascular surgery with cardiopulmonary bypass. The inclusion criteria were adult age 18 to 80 years that underwent myocardial revascularization, valve replacement and combined surgeries in moderate hypothermia (27° C - 32° C). The research excluded patients submitted to reoperation, surgical correction of congenital heart diseases, circulatory arrest and cardiac transplantation.

Patients were divided in two groups, each group containing 20 individuals. For both groups, the technique used during the cardiopulmonary bypass was taken into consideration. Group 1 was composed of patients who would undergo surgical procedures with CPB using blood gas analysis at intermittent intervals (20 - 30 minutes). Group 2 was specific to patients that underwent surgical procedures with a capnography system adapted to the membranes oxygenator exhaustion and blood gas test.

All the patients received similar perioperative care; the CPB circuit was prepared with 2000 ml of 0.9% sodium chloride, 100 ml of human albumin and 50 ml of 8.4% sodium bicarbonate, the heparinization was performed at a dose of 4 mg / kg, the start of cardiopulmonary bypass occurred when the activated coagulation time (ACT) exceeded values over 480 seconds. After CPB initiation, stabilization of the total flow (40ml/kg/min - 60ml/kg/min) and cessation of lung ventilation, a PVC tube was connected to the outlet of the CPB circuit, being sequentially adapted to a capnograph sensor connected to the Dräger Primus® anesthesia machine, the data was digitized and transferred to the monitor of the device, being possible to monitor the oxygenator gas exhaustion in real time (Figure 1). Continuous measurement of oxygenator PCO_2 was performed based on reference values between 35mmHg - 45mmHg, which are the parameters related to arterial PCO2. The PH-stat technique was used during the CPB procedure and the adjustment of the carbon dioxide

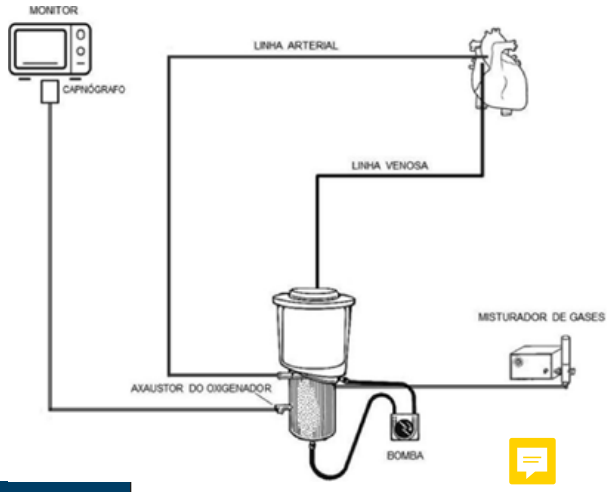

Figure 1 *Visão geral esquemática da configuração do capnógrafo adaptado ao oxigenador de membranas.*

pressure was done according to the strategies used by the perfusionist. Blood pressure was maintained at values of 50 to 70 mmHg.

Arterial blood samples were collected for blood gas test using a GEM® Premier™ 4000 automatic device, which measured the concentration the parameters. The temperature of the sample was corrected according to the patient esophageal temperature.

Three brands of membrane oxygenator were used: Medtronic Affinity, Sorin Inspire 6F / 8F and Nipro Vital, operated combined with continuous flow centrifugal pumps.

In group 1, the blood samples were collected for the blood gas analysis and obtained through an arterial line of the cardiopulmonary bypass circuit and then sent to laboratory analysis. Simultaneously, we recorded esophageal temperature.

Group 2 continuous monitoring of CO_2 partial pressure of membrane oxygenator exhaustion ($PeCO_2$) was observed through the capnographer adapted to the CPB. Blood samples were collected for the arterial blood gas test, and we recorded the values presented by the capnography and the esophageal temperature.

The average and standard deviation of the data collected from both groups were obtained. T-test was used to compare *arterial partial pressure* of carbon dioxide ($PaCO_2$) in group 1 and group 2. The value $p < 0.05$ was considered statistical significance. In group 2 the correlation between $PeCO_2$ and $PaCO_2$ were evaluated by linear regression. The Bland-Altman method was used to determine the degree of agreement between the two variables.

RESULTS

25 male patients and 15 female, age 41-79 years (av. 61,6 years) participated in this study. 83 blood samples were collected and analyzed. The esophageal temperature varied between 28º and 32,9º C (av. 32,1º C). The cardiopulmonary bypass time varied from 43 to 205 minutes (av. 96 minutes) and the anoxia time, from 36 to 185 minutes (av. 53 minutes). Table 1 shows the average and standard deviation of group 1's PaCO2 and group 2's $PaCO_2$ / $PeCO_2$.

For the conventional criteria, the difference between group 1 and group 2 is considered not statistically significant ($P = 0.21$). The average $PaCO_2$ in both groups was not significantly different, and these data are presented in table 2.

Table 1 **Group 1 and Group 2 - Average ± SD (mmHg)**

	n	Média ± SD
Group 1's $PaCo^2$	42	44%
Group 2's $PaCo^2$	41	36.5 ± 4.42
$PeCo^2$		39.9 ± 3.98

$PaCO_2$ - arterial CO_2 partial pressure (mmHg); $PeCO_2$ - CO_2 partial pressure of oxygenator exhaust (mmHg)

Table 2 **Average ± SD (mmHg) $PaCO_2$**

	Group 1	Group 2	*p*
$PaCO_2$ (Max/Min)	34.6±7.44 (20/53)	36.5 ± 4.42 (27/48)	0,21

$PaCO_2$ - arterial CO_2 partial pressure (mmHg); P - T-test comparing Group 1 and Group 2. (P <0.05)

Figure 2 demonstrates the correlation between $PaCO_2$ and $PeCO_2$ measured in group 2. The temperature of the sample was corrected according to the esophageal temperature of the patients and showed a significant positive correlation (r = 0.79, p <0.001). Figure 3 presents the graphical analysis of Bland-Altman between the values of PaCO2 corrected from esophageal temperature and the PeCO2. It's possible to verify an average agreement of difference between the results corresponding to 3.47 ± 2.70 mmHg. The 95% upper and lower limits (dashed lines) correspond to -1.8 and 8.76 mmHg, respectively.

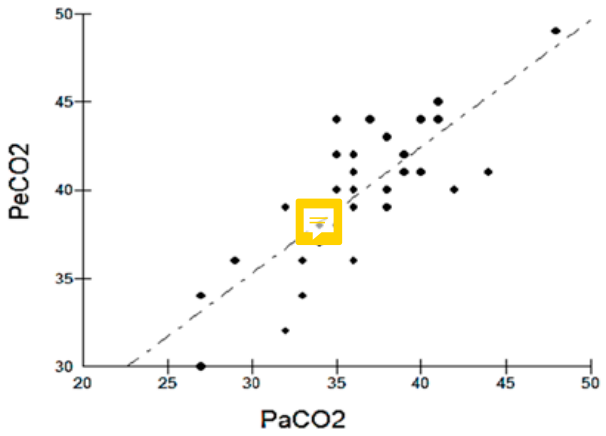

Figure 2 *Correlação entre a pressão parcial de CO2 do escape do oxigenador de membranas (PeCO2) e a pressão parcial de CO2 arterial (PaCO2). r = 0,79, P < 0,001.*

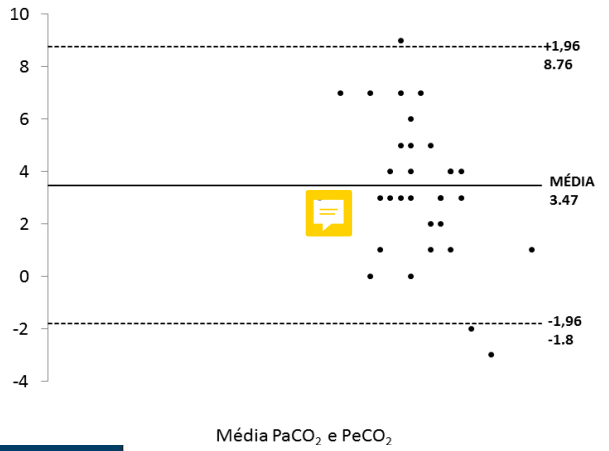

Figure 3 *Concordância entre a PeCO2 (Escape do oxigenador) e o PaCO2 (Arterial) em hipotermia moderada durante a circulação extracorpórea.*

DISCUSSION

This study demonstrates that there was no statistically significant difference in the $PaCO_2$ measurements in both groups; we observed that both groups contain relatively equal average statistically. The results show that the capnography of the oxygenator provides a level of accuracy comparable to the conventional analysis of the blood gas test. Although the $PaCO_2$ of group 1 and group 2 show statistical similarity, the continuous monitoring of PCO_2 of the oxygenator exhaust provides a faster evaluation, which can avoid unwanted outcomes to the patient. Graham *et al*.[4] indicate in their studies that membrane oxygenator capnography is a real-time indicator for estimating arterial PCO_2. This device causes a safe and proper elimination of carbon dioxide during the cardiopulmonary bypass. However, despite the favorable results obtained in the research, Graham *et al*.[4] clarify that this instrument should not replace the intermittent collections of blood gases during the procedure.

The results of this study indicate that the analyzed $PeCO_2$ and $PaCO_2$ corrected from esophageal temperature obtained a positive linear correlation (r = 0.79, P <0.001) in group 2. These results come to agreement to the reports of Potger *et al*.[5] (r = 0.83 P <0.001), in which a strong positive correlation was observed between PCO_2 of the membrane oxygenator exhaustion and the arterial carbon dioxide. $PeCO_2$ measured by the capnographer adapted to the membrane oxygenator presented values above the average $PaCO_2$. However, the degree of agreement was with an average of (3.47 ± 2.70 mmHg). Baraka *et al*.[6] reported an average of (2.8 ± 2.0 mmHg) to express agreement in the moderate hypothermia. Although these results present different values from those presented in our study, both studies are strictly in concord. This difference can be attributed to the use of a thermometer connected directly to the CPB circuit, which demonstrated the arterial blood temperature measurement, while we used the esophageal temperature to make the correction in the arterial blood gas analysis in our study. Baraka *et al*.[6] also describe that the continuous measurement of PCO_2 by capnography is a reliable instrument in normothermia and stable hypothermia during CPB. However, they demonstrate that during hypothermia one should take into account the correction of the temperature of the sample to obtain great results, in this case the PH-stat strategy is the method used during the procedure.

In group 2, the results presented $PeCO_2$ average higher than those of $PaCO_2$, and we can assume that maintaining the CO_2 of capnography in overestimated values preserve a possible stability in arterial PCO_2 results. This observation agrees with Potger *et al*.[5] that identified the capnography overestimate the PCO_2 on some occasions during cardiopulmonary bypass. However, this difference was not considered significant. In addition, this disparity is clinically acceptable and does not interfere with the use of capnography.

The oxygenators; Medtronic Affinity, Sorin Inspire 6F / 8F and Nipro Vital were used in a random way, attesting

that all of them presented a strong concordance and correlation between $PeCO_2$ and $PaCO_2$ in group 2, independent of the oxygenator used for the measurement of carbon dioxide. Previous researches have used only one oxygenator brand to validate the study, which would raise doubt about the effectiveness of capnography. The associate evaluation of several models in one single study demonstrates that the $PeCO_2$ values are totally dependent on the technique that is, the membrane type does not interfere with the results presented in the capnographer. These findings emphasize and confirm that the professional perfusionist has the responsibility to monitor and use strategies to maintain CO_2 levels within the appropriate physiological parameters.

CONCLUSION

It is concluded that continuous PCO_2 monitoring of the membrane oxygenator exhaustion of the cardiopulmonary bypass circuit has a positive impact on the CO_2 arterial blood gas test results. This instrument confirms and maintains the carbon dioxide values into the reference parameters, as well as being a lower cost option, which makes it accessible in services where the use of in-line blood gas monitoring is not part of the routine.

REFERENCES

1. Pereira IB, Batista DCS. O perfusionista e o equilíbrio ácido/base durante a circulação extracorpórea. Rev Elet Acer Sau. 2013; 5(2): 456-473.
2. Luz HLM, Auler Júnior JOC. Temperature and Acid-Base Balance in Coronary Bypass Grafting with Cardiopulmonary Bypass, under Hypothermia and Normothermia. Rev Bras Anestesiol. 2002; 52(2):197-208.
3. Pereira M, Vilela H, Pina L. Capnografia como método de monitorização ventilatória durante estados de sedação induzida. Rev Soc Port Anestesiol. 2006; 14(4): 24-28.
4. Graham JM, Gibbs NM, Weightman WM, Sheminant MR. The relationship between oxygenator exhaust PCO2 and arterial PCO2 during hypothermic cardiopulmonary bypass. Anaesth Intensive Care. 2005; 33(4): 457–461.
5. Potger KC , McMillan D , Southwell J , Dando H , O'Shaughnessy K. Membrane oxygenator exhaust capnography for continously estimating arterial carbon dioxide tension during cardiopulmonary bypass . J Extra Corpor Technol. 2003; 35(3): 218-223.
6. Baraka A, El-Khatib M, Muallem E, Jamal S, Haroun-Bizri S, Aouad M. Oxygenator exhaust capnography for prediction of arterial carbon dioxide tension during hypothermic cardiopulmonary bypass. J Extra Corpor Technol. 2005; 37(2): 192–195.
7. Abreu TT. Análise da variação de eletrólitos após cirurgia cardíaca com uso de circulação extracorpórea [Monografia]. Salvador: Universidade Federal da Bahia – UFBA; 2014.
8. Antunes N, Dragosavac D, Petrucci O, Oliveira M, Kosour C, Blotta M, Braile M, Vieira R. The use of ultrafiltration for inflammatory mediators removal during cardiopulmonary bypass in coronary artery bypass graf surgery. Braz J Cardiovasc Surg. 2008;23(2):175-82.
9. Guyton AC, Hall JE. Tratado de fisiologia médica. 12ª ed. Rio de Janeiro: Elsevier; 2011.
10. Kristiansen F, Hogetveit JO, Pedersen TH. Clinical evaluation of an instrument to measure carbon dioxide tension at the oxygenator gas outlet in cardiopulmonary bypass. Perfusion. 2006; 21(1): 21–26.
11. Madias NE. Renal acidification responses to respiratory acid--base disorders. J Nephrol. 2010; 23(16): 85–91.
12. Mota AL, Rodrigues AJ, Évora PRB. Adult cardiopulmonary bypass in the twentieth century: science, art or empiricism?? Braz J Cardiovasc Surg. 2008; 23(1): 79-92.
13. Souza MHL, Elias DO. Fundamentos da Circulação Extracorpórea. 2ª ed. Rio de Janeiro: Alfa Rio; 2006.
14. Torrati FG, Dantas, RAS. Circulação extracorpórea e complicações no período pós-operatório imediato de cirurgias cardíacas. Acta Paul Enferm. 2012; 25(3): 40-345.
15. Taniguchi FP, Souza AR, Martins AS. Tempo de circulação extracorpórea como fator risco para insuficiência renal aguda. Rev Bras Cir Cardiovasc. 2007; 22(2): 201-5.

ARTIGO ORIGINAL ORIGINAL ARTICLE

ANEURISMAS VERDADEIROS DO MEMBRO SUPERIOR: REVISÃO DA EXPERIÊNCIA DE UM SERVIÇO

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Resumo

Introdução: Os aneurismas verdadeiros do membro superior são raros e o seu tratamento visa a prevenção de complicações como a embolização distal, a compressão de estruturas neurovasculares adjacentes ou a rotura.

Objetivo: O objetivo deste estudo é rever a casuística de um serviço no tratamento cirúrgico de aneurismas verdadeiros do membro superior.

Métodos: Foi realizado um estudo retrospetivo entre Janeiro de 2007 e Agosto de 2017.

Resultados: De um total de onze doentes, nove eram do sexo masculino e dois do sexo feminino. Um dos doentes foi submetido a duas cirurgias por aneurismas consecutivos do membro superior. De um total de doze casos, dois tinham localização na artéria subclávia, um na axilar e nove na braquial. Três aneurismas eram de etiologia degenerativa/idiopática, um estava associado à presença de costela cervical e sete ocorreram no contexto de fístula arteriovenosa para hemodiálise e/ou transplante renal. Cinco doentes foram submetidos a cirurgia em contexto de urgência e os restantes em contexto eletivo. Todos os pacientes foram submetidos a aneurismectomia. A morbilidade aos 30 dias correspondeu a dois hematomas, um síndrome do compartimento e duas oclusões precoces com um total de quatro pacientes a necessitarem de reintervenção. Durante o período de *follow-up* todos os doentes com o enxerto inicialmente preservado apresentaram permeabilidade dos enxertos. Não houve necessidade de cirurgia mutiladora.

Conclusão: Neste estudo, a maioria dos aneurismas do membro superior ocorreram em doentes com fistulas arteriovenosas para hemodiálise e/ou transplantados renais. Apesar da necessidade de reintervenção em alguns casos, o tratamento cirúrgico dos aneurismas do membro superior acarreta uma baixa morbilidade.

Abstract

True aneurysms of the upper limb: a single-centre experience

Introduction: *True arterial aneurysms of the upper limb are rare and their treatment is intended to avoid complications as distal embolization, compression of surrounding neurovascular structures or rupture.*

Objective: *The purpose of this study is to review the experience of a department in the surgical treatment of true arterial aneurysms of the upper limb.*

Methods: *A retrospective study was performed between January 2007 and August 2017.*

Results: *From a total of eleven patients, nine were male and two were female. One of the patients had surgery twice because of two consecutive aneurysms of the upper limb. From a total of twelve cases, two were subclavian, one was axillary and nine were brachial aneurysms. Three of them had degenerative/idiopathic aetiology, one was associated to a cervical rib and seven occurred in the setting of arteriovenous fistula or kidney graft. Five patients had emergent surgery and the others had elective surgery. All of the patients were submitted to aneurysmectomy. As 30-day complications, there were two haematomas, one compartment syndrome and two early graft occlusions. Four patients needed re-intervention. During the follow-up period, all the grafts initially preserved were patent. There were no further known complications or amputation procedures.*

Conclusion: *In this review most of the aneurysms were found in patients with haemodialysis vascular access or kidney grafts. Despite the need for early re-intervention in some cases, the surgical treatment of true arterial aneurysms of the upper limb is a low morbidity procedure.*

INTRODUÇÃO

Os aneurismas verdadeiros do membro superior são raros. Mais do que evitar a rotura, o seu tratamento visa a prevenção de complicações mais frequentes como a embolização distal ou a compressão de estruturas neurovasculares adjacentes.

Os aneurismas do membro superior podem ser degenerativos, estar associados a alterações congénitas, nomeadamente do tecido conjuntivo, ou surgirem em contexto de traumatismo. No entanto, e consoante a localização, podem estar associados a outras etiologias. Os aneurismas subclávios e axilares podem estar associados à presença de uma costela cervical/síndrome do desfiladeiro torácico ou estar relacionados com causa infecciosa (sífilis, tuberculose). Por outro lado, os aneurismas da artéria braquial são mais frequentes em doentes com acessos vasculares para hemodiálise e/ou transplantados renais, tendo esta relação sido previamente documentada.[1,2]

Independentemente do contexto etiológico, está recomendada a correção de aneurismas do membro superior. A cirurgia convencional continua a desempenhar um papel preponderante no tratamento destes doentes e está associada a excelentes resultados.[1,3]

O objetivo deste estudo é rever a casuística de um Serviço de Angiologia e Cirurgia Vascular no tratamento cirúrgico de aneurismas verdadeiros do membro superior.

MÉTODOS

Neste trabalho foi realizada uma análise retrospetiva dos doentes com aneurismas arteriais verdadeiros do membro superior, submetidos a cirurgia, entre 1 de Janeiro de 2007 e 1 de Julho de 2018 no Serviço de Angiologia e Cirurgia Vascular do Centro Hospitalar e Universitário de Coimbra. Foram identificados doze casos correspondentes a onze doentes. Os dados foram recolhidos através da consulta do processo clínico dos doentes e foram colhidas informações sobre o sexo, idade, antecedentes médicos, etiologia do aneurisma, procedimento cirúrgico, complicações associadas, necessidade de reintervenção e *follow-up*.

RESULTADOS

De um total de onze doentes submetidos a cirurgia por aneurismas arteriais verdadeiros do membro superior, nove doentes eram do sexo masculino e dois do sexo feminino. As idades dos doentes estavam compreendidas entre os 29 e os 68 anos (idade média de 51,4). Um dos doentes apresentou dois aneurismas consecutivos da artéria braquial e foi submetido a dois procedimentos cirúrgicos, contribuindo para um total de doze casos analisados. Os antecedentes médicos de cada doente encontram-se discriminados na tabela 1. É de realçar que sete dos onze doentes considerados (64%) têm antecedentes de fístula arteriovenosa (FAV) para hemodiálise e que seis desses doentes (55%) foram submetidos a um transplante renal e apresentavam a fístula funcionante à data da cirurgia. O único caso que mantinha necessidade de hemodiálise esta foi realizada através da colocação de um cateter venoso central. Relativamente ao tipo de fístula arteriovenosa, um doente tinha uma fístula do tipo radiocefálica, dois doentes tinham fístulas rádio e umerocefálicas e quatro doentes tinham apenas fístulas umerocefálicas. Todos os doentes submetidos a transplante renal apresentavam o enxerto funcionante à data do estudo.

Tabela 1 **Antecedentes médicos**

Antecedentes médicos	Nº de doentes
Dislipidemia	5
Hipertensão Arterial	6
Diabetes Mellitus	2
Tabagismo	1
Outros aneurismas	1
Fístula arteriovenosa para hemodiálise	7
Transplante renal	6
VHC + VIH	3

VHC: Vírus da Hepatite C; VIH: Vírus da Imunodeficiência Humana

De um total de doze casos, dois doentes tinham aneurismas da artéria subclávia. Um dos aneurismas era de provável etiologia degenerativa e o outro encontrava-se associado à presença de costela cervical. No último caso o doente foi operado em contexto de urgência por embolização distal. Outro caso foi o de um aneurisma axilar de provável etiologia degenerativa num doente com múltiplos fatores de risco ateroscleróticos e com um aneurisma concomitante da aorta torácica descendente. Os outros nove casos eram de aneurismas da artéria braquial, no contexto de FAV/hemodiálise ou transplante renal, à exceção de um caso que era de etiologia idiopática.

Todos os casos foram submetidos a aneurismectomia, total ou parcial. Foi realizada uma anastomose topo-a topo em três casos, interposição de enxerto em oito casos (um caso com enxerto venoso e sete casos com PTFE) e endoaneurismorrafia em um caso.

Cinco casos foram operados em contexto de urgência, dos quais três em contexto de isquémia aguda por trombose/embolização distal do aneurisma e dois em contexto de rotura com fistulização cutânea. Os sete casos operados em contexto eletivo eram na maioria assintomáticos (cinco casos). Os doentes sintomáticos apresentavam dor, parestesias e/ou sensação de arrefecimento na mão. Cada caso encontra-se detalhadamente descrito na tabela 2.

Foram analisadas as complicações com necessidade de reintervenção aos 30 dias. A complicação mais frequente foi oclusão do enxerto em 17% dos casos, com posterior realização de trombectomia do enxerto. Em 8% dos casos ocorreu síndrome do compartimento com necessidade de fasciotomia e hematoma com necessidade de drenagem cirúrgica. As complicações e abordagem de cada caso estão descritas na tabela 3. Um dos doentes que foi operado em

Tabela 2 **Caracterização dos casos**

Caso	Idade	Local do aneurisma	Etiologia	Contexto da cirurgia	Antecedentes
Caso 1 M	29	Subclávio	Costela cervical	Urgência (embolização distal)	-
Caso 2 M	68	Subclávio	Degenerativo	Eletiva	Dislipidemia, HTA
Caso 3 M	64	Axilar	Degenerativo	Eletiva	HTA, tabagismo, outro aneurisma, DM, Síndrome nefrótico, Miocardiopatia hipertrófica
Caso 4 M	41	Braquial	FAV / TR	Eletiva	Dislipidemia, VHB/VHC
Caso 5 M	42	Braquial	FAV / TR	Eletiva	Dislipidemia, VHB/VHC
Caso 6 F	55	Braquial	FAV / TR	Urgência (rotura)	Dislipidemia, FA, Valvuloplastia mitral
Caso 7 M	46	Braquial	FAV	Eletiva	VHB/VHC, doença coronária
Caso 8 M	45	Braquial	FAV / TR	Eletiva	VHB/VHC, Dislipidemia, HTA
Caso 9 F	63	Braquial	FAV / TR	Eletiva	Dislipidemia, HTA, DM
Caso 10 M	47	Braquial	Idiopático	Urgência (trombose)	-
Caso 11 M	57	Braquial	FAV / TR	Urgência (rotura)	HTA
Caso 12 M	60	Braquial	FAV / TR	Urgência (embolização distal)	HTA

M- Masculino; F- Feminino; HTA- Hipertensão; DM- Diabetes Mellitus; FA- Fibrilhação auricular; VHB- Vírus da Hepatite B; VHC- Vírus da Hepatite C; TR- Transplante renal

contexto de urgência por rotura de aneurisma da artéria braquial (caso 6) apresentou no pós-operatório precoce um hematoma e oclusão precoce do enxerto com necessidade de drenagem e trombectomia do enxerto. Tendo em conta o grau de perda tecidular decorrente de duas abordagens cirúrgicas foi difícil o controlo da hemostase durante o período pós-operatório apesar das medidas conservadoras instituídas. O doente foi submetido posteriormente a exérese do enxerto por re-trombose e laqueação da artéria axilar. Um doente com oclusão do enxerto no pós-operatório precoce foi submetido a trombectomia (caso 8). Um dos casos operados em contexto de isquemia aguda por trombose de aneurisma da artéria braquial apresentou síndrome compartimental com necessidade de fasciotomia que foi realizada em segundo tempo (caso 10). Nos casos analisados não foram realizadas cirurgias mutiladoras nem se verificou mortalidade.

No período de *follow-up* (mediana de 11 meses), e em todos os casos com preservação inicial do enxerto (exceto o caso 6), os enxertos mantiveram-se patentes. O caso que cursou com laqueação da artéria axilar (caso 6) apresentou evolução lenta da cicatrização da ferida no pós-operatório, no entanto manteve-se assintomática durante o follow-up. Um dos doentes (caso 10 da tabela 3) que foi operado em contexto de urgência por trombose de aneurisma com isquémia aguda do membro superior e necessidade de posterior fasciotomia por síndrome do compartimento manteve queixas de parestesias e diminuição da força muscular digital durante o período de *follow-up*.

Tabela 3 **Complicações**

Caso	Complicações	1ª reintervenção	2ª reintervenção
3	Hematoma	Drenagem	-
6	Hemorragia e oclusão do enxerto	Drenagem e trombectomia	Exérese do enxerto e laqueação da artéria axilar
8	Oclusão do enxerto	Trombectomia	-
10	Síndrome do compartimento	Fasciotomia	Desbridamento cirúrgico

DISCUSSÃO

Os aneurismas verdadeiros do membro superior são raros. Nesta revisão, a maioria dos aneurismas dos membros superiores são aneurismas da artéria braquial em pacientes com FAV para hemodiálise e enxerto renal concomitante. Esta associação já tem sido descrita em estudos previamente publicados.

Vários fatores podem contribuir para o desenvolvimento de aneurisma da artéria braquial nestes doentes. Um dos fatores julga-se estar relacionado com o padrão de fluxo hiperdinâmico na artéria proximalmente à FAV que cursa com alterações ao nível da parede arterial e com uma maior propensão à sua degenerescência aneurismática.[4] Ainda, verificou-se que esta suscetibilidade permanece mesmo após a laqueação da FAV sugerindo que por si só a laqueação da FAV não altera o curso da doença e que outros fatores poderão estar subjacentes a este fenómeno.[5] Um deles é a corticoterapia realizada nos doentes transplantados que através do *remodelling* da parede arterial também pode influenciar a degenerescência aneurismática nestes doentes.[6]

Neste estudo, apesar da necessidade de reintervenção em alguns casos, o tratamento cirúrgico convencional dos aneurismas do membro superior revelou-se um procedimento associado a uma baixa morbilidade e com elevada permeabilidade do enxerto. Estes resultados vão de encontro aos resultados previamente publicados, quer no que diz respeito à morbilidade dos doentes, quer no que diz respeito à permeabilidade do enxerto, com taxas de permeabilidade a rondar os 100%.[1,2,3] Em relação ao tipo de conduto, os condutos venosos apresentam uma permeabilidade superior aos protésicos devendo, por isso, ser o conduto de eleição.[7,8] Contudo, os doentes podem apresentar falência do acesso o que leva à necessidade de construi um novo acesso e ao eventual esgotamento futuro de conduto venoso disponível.[9] Neste trabalho a maioria dos doentes foi submetida a revascularização com conduto protésico por ausência de capital venoso ou de veias alvo adequadas para a realização do acesso.

Os aneurismas verdadeiros dos membros superiores são raros. A evidência existente ao nível da literatura limita-se a estudos com amostras reduzidas o que dificulta a validação dos resultados no que diz respeito ao tratamento ideal desta patologia quer, por exemplo, no que toca à inferência de eventual vantagem na adopção de programas de vigilância destes doentes, principalmente daqueles com aneurismas associados a FAV e transplante renal. Apesar da necessidade de mais estudos que abordem esta patologia, o tratamento cirúrgico convencional dos aneurismas do membro superior continua a ser o procedimento de escolha na abordagem destes doentes.

REFERÊNCIAS

1. Chemla E, Nortley M, Morsy M. Brachial artery aneurysms associated with arteriovenous access for hemodialysis. Semin Dial 2010; 23:440-444.
2. Marzelle J, Gashi V, Nguyen HD, Mouton A, Becquemin JP, Bourquelot P. Aneurysmal degeneration of the donor artery after vascular access. J Vasc Surg 2012; 55:1052-1057.
3. Tetik O, Yilik L, Besir Y, Can A, Ozbek C, Akcay A, Gurbuz A. Surgical treatment of axillary artery aneurysm. Tex Heart Inst J 2005; 32:18-188.
4. Graham JM, McCollum CH, Crawford ES, ME DeBakey. Extensive arterial aneurysm formation proximal to ligated arteriovenous fistula. Ann Surg. 1980;191:200-2.
5. Anastasiadou C, Megalopoulos A, Tasiopoulou K, Intzos V. A Rare Case of Brachial Artery Aneurysm Following Hemodialysis Fistula Ligation in a Transplanted Patient. Vasc Endovascular Surg. 2019 Jan;53(1):71-74.
6. Martins P, Ferreira T, Manuel V, Tiago J, Silvestre L, Fernandes R, Pedro LM, Nunes JS, Fernandes JF. Aneurisma umeral em doentes com transplante renal. Angiologia e Cirurgia Vascular 2014; 10:58-63
7. McCarthy WJ, Flinn WR, Yao JS, Williams LR, Bergan JJ. Result of bypass grafting for upper limb ischemia. J Vasc Surg 1986; 3:741-746.
8. Huber TS, Carter JW, Carter RL, Seeger JM. Patency of autogenous and polytetrafluoroethylene upper extremity arteriovenous hemodialysis accesses: a systematic review. J Vasc Surg. 2003 Nov;38(5):1005-11.
9. Tushar J. Vachharajani, Anil K. Agarwal, Arif Asif. Vascular access of last resort. Kidney International (2018) 93, 797–802.

ARTIGO DE REVISÃO REVIEW ARTICLE

NEOINTIMAL HYPERPLASIA

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Abstract

Neointimal hyperplasia is a physiologic healing response to injury to the blood vessel wall, involving all the three arterial layers and it occurs in the presence of internal (endovascular) or external (surgical) injury.

It is a highly complex process involving several tissues (perivascular, vessel wall, and blood) and numerous cell lineages with multiple molecular signaling networks. So, there is a number of possible targets for inhibition of this process. There are known risk factors for Intimal Hyperplasia, such as diabetes, female gender, presence of systemic inflammation, type of arteries treated, types of surgical and endovascular materials, presence of turbulent flow and genetic status.

The present paper discusses the pathophysiology of neointimal hyperplasia and the strategies to prevention and treatment of it.

INTRODUCTION

In vascular surgery, just like in our lives, everything we do has consequences. Newton said so when he first described his third laws of motion, published in 1687. The translation of this principle for vascular surgery is called neointimal hyperplasia. Neointimal hyperplasia is a physiologic healing response to injury to the blood vessel wall, in a process analogous to scar formation.

When there is an aggression (surgery, angioplasty, stenting, altered flow pattern), endothelium stimulation occurs, with platelet activation and aggregation and leucocyte adhesion and infiltration – this is the beginning of the thrombotic and inflammatory process. At the site of injury, endothelial cells are denuded, and the subendothelial matrix is exposed to flowing blood. Platelets and fibrinogen immediately adhere to the surface of the injured vessel. A multistep cascade of platelet and leukocyte adhesion and activation promotes secretion of inflammatory signaling molecules and growth factors that promote smooth muscle cells migration from the media to the intima.[1] The smooth muscle cells proliferate in the intima and deposit extracellular matrix, release degrading enzymes with basement membrane destruction and then transmigrate into the extraluminal tissue, causing stenosis.[1]

The adventitia plays a critical role in this process. The adventitia is considered an "injury sensing tissue", due to its capability to respond to different stimuli in an outside-in manner, toward the intima and leading to vessel remodeling. Adventitial fibroblasts when stimulated, undergo phenotypical changes to myofibroblasts, which are specialized cells with contractile as well as migratory properties. Myofibroblasts can undergo proliferation and migration to form neointimal tissue.

All the three arterial layers are involved in the pathophysiology of neointimal hyperplasia and why it occurs in the presence of internal (endovascular) or external (surgical) injury. It is a highly complex process involving several tissues (perivascular, vessel wall, and blood) and numerous cell lineages with multiple molecular signaling networks. So, there are an amount of possible targets for inhibition of this process, currently under investigation.

The aim or this review is to give insight into the process of intimal hyperplasia focusing on risk factors and strategies developed so far to fight against in vascular intervention.

RISK FACTORS

There are known risk factors for Intimal Hyperplasia, such as diabetes, female gender, presence of systemic inflammation, type of arteries treated, surgical and endovascular materials, presence of turbulent flow and genetic status (IL-10 gene, chromosome 12). Diabetes is associated with increased endothelial dysfunction, increased platelet activity and more aggressive cellular response to injury. Female patients generally have smaller vessels and there may be some hormone contribution to their higher risk of development of intimal hyperplasia. Systemic inflammation, measured with C reactive protein, lipoprotein (a), postprocedural von Willebrand factor, and plasminogen activator inhibitor-1 antigen also correlate with unfavorable outcomes. Muscular (distributing) arteries, with high

vascular smooth muscle content in their media have higher restenosis rate than elastic (conductance) arteries. Iliac arteries are conductance vessels (elastic) with a high elastin content in their media. Consequently, the rate of restenosis is expected to be relatively low. Profunda femoral, popliteal, and tibial arteries are muscular (distributing) arteries, with high rates of restenosis. One of the most powerful predictors of restenosis is the vessel diameter, as we see that smaller vessels are at greater risk of restenosis. Stents and prosthetic grafts cause more inflammatory stimulus, with consequently more restenosis due to intimal hyperplasia.

Hemodynamic forces, specifically shear stress and wall tensile stress, are well-established initiators and modulators of intimal hyperplasia. Under physiologic conditions, the steady laminar blood flow generates shear stress in arteries. Endothelial cells sense this physiologic shear force, releasing mediators such as nitric oxide (NO) maintain a quiescent state for smooth muscle cells and homeostasis of the whole vessel wall.[2,3] Vascular reconstructions such as vein bypass grafts, stented diseased arteries, and arteriovenous fistulas not only alter the rate of the local blood flow but also frequently induce a disordered flow pattern. In normal and increased wall shear stress protective pathways are induced, particularly anti-inflammatory and antioxidant pathways.[3] This explains how an exercise training program is specially effective in claudicants.[3] Both clinical and experimental observations have demonstrated that disturbed flow and/or low wall shear stress accelerate development of intimal hyperplasia. Endothelial cells respond to these particular hemodynamic conditions by elaborating adhesion molecules and proinflammatory cytokines that in turn enhance cell proliferation and matrix accumulation, leading to robust intimal growth.[4] On the other hand, laminar, high blood flow (uniform high shear stress) generally exhibits an opposing effect on intimal growth. Furthermore, augmentation of blood flow in vessels with established intimal hyperplasia induces intimal regression. This knowledge has been translated to clinical application, so that creation of a distal fistula to boost the blood flow has led to improvement in the patency rate of lower extremity grafts in selected circumstances. We know the nature of the flow through the vessel will have a major effect. High flow resulting in high wall shear stress in a unidirectional uniform laminar pattern will push the vessel towards maturation or healthy re-modelling, whereas low wall shear stress with chaotic turbulent and oscillating flow will encourage the vessel to remodel inwards.[2]

TREATMENT

So how can this knowledge be used in terms prevention and treatment of intimal hyperplasia? We can prevent intimal hyperplasia by limiting the extent of endothelial injury, minimizing trauma to the adjacent normal vessel, with selective use of stents and prosthetic devices and gentle and careful handling of tissues (particularly the adventitia) during surgical procedures and during endovascular procedures. We can induce healthy flow patterns with grafts and avoid pathological flow, using adequate graft designs and anastomotic angles. Also, there are some drugs that claim to improve overall flow. We are yet to develop an agent which makes the endothelium more protected to these stimuli, to switch the endothelial cells to atheroprotective phenotype, witch makes this is an important area for study. Attempts have been made to block the effect of factors on the cells with antiproliferative agents and more excitingly with allogeneic cells.

Remembering the process of neointimal hyperplasia formation, we can inhibit each step of the cascade with endothelial protective agents, antiplatelet therapy, anticoagulants, anti-inflammatory drugs, antiproliferative agents and by the use of radiation and brachytherapy. In what concerns blockade of the cellular response to the signals, these are some of the possible targets for inhibition of intimal hyperplasia under investigation, such as selectins, integrins, ICAM, VCAM; interleukins,[1,6,9,10,19] TNF-β, MMP[2,9] and the Renin-Angiotensin System. Biologic therapy can be delivered via three platforms: drug-eluting stents (DES), drug-coated balloons (DCB) and via direct drug delivery. DES and DCB normally use drugs as paclitaxel and sirolimus, which are immunosuppressants, antiproliferative, anti-inflammatory and antimitotic drugs. Paclitaxel binds to the beta subunit of tubulin, leading to the inhibition of microtubule disassembly. By blocking microtubule disassembly, paclitaxel prevents cell progression from G2 to M, interferes with the mitotic spindle apparatus, inhibits smooth muscle cell migration, and signal transduction. Sirolimus is a weak antibiotic but a powerful immunosuppressant and antimitotic. It blocks the cell cycle from progressing from G1 to the S phase.

Drug-eluting stents

DES are excellent therapies in vessels with significant elastic recoil and/or flow limiting dissections. They improved primary patency and reduced target lesion revascularization when compared to balloon angioplasty for femoropopliteal lesions as demonstrated by some studies as the ZILVER PTX (primary patency of 66.4% with Zilver PTX DES compared to 43.4% with PTA alone at 5 years)[5] and the MAJESTIC Trial (primary patency of 83.5% with Eluvia DES at 3 years).[6] DES major disadvantages are that they lose their anti-neoproliferative over time and become a bare stent which may elicit a long-term inflammatory response and predispose a patient to develop restenosis of the vessel segment. Also, DES impairs re-endothelization and exacerbate life-threatening stent thrombosis because of endothelium damage caused by both drug and stenting.

Drug-coated balloons

DCB overcome the limitation of leaving a permanent prosthesis. They have also shown beneficial long-term outcomes above the knee compared to PTA alone as demonstrated by the IN.PACT SFA (primary patency of 82.2% vs. 52.4% with PTA alone at 1 year),[7] the Lutonix

Trial (24% Target lesion revascularization rate at 2 years)[8] and the ILLUMENATE study (primary patency of 76.3% for DCB compared to 57.6% for PTA alone).[9]

Drug coated balloons (DCB) designed to release antimitogenic agents to the site of the blockage are aimed at reducing artery restenosis after intervention. However, first generation DCB utilize mainly direct application of the chemotherapy drugs along with hydrophilic excipients to facilitate uptake into the tissue, and the majority of drug is released from the DCB systemically. Unfortunately, only a smaller fraction (~20%) of the anti-neoproliferative on the balloon is delivered to the vessel wall and the downstream shedding of anti-neoproliferative could be deleterious.

Recently, some concern has been raised with a positive mortality signal related to the use of paclitaxel.[10] In the three RCTs composed of 863 patients with longer-term follow-up of 4 years (IN.PACT SFA, presented but not yet published) and 5 years (Zilver PTX, THUNDER), the all-cause death increased further with paclitaxel (14.7% vs 8.1%, crude risk of death; RR, 1.93; 95% CI, 1.27–2.93; number needed to harm, 14 patients [95% CI, 9–32]). The absolute risk difference was 7.2% (95% CI, 3.1–11.3%). There was no statistically significant heterogeneity between studies (P = .92).[10]

There is limited data for the use of DCB below the knee. The IN.PACT DEEP trial was stopped early because it did not meet its primary endpoint and there were signals suggestive of an increased rate of major amputation in the DCB arm.[11] The BIOLUX study demonstrated similar primary patency loss between PTA and DCB (17.1% patency loss in DCB vs. 26.1% in PTA, p =0.298).[8]

An important problems related to the use of DES and DCB is that there we are dozens of sponsored trials, all of them with excellent procedure success rates, measured as TLR (target lesion revascularization), but with no differences in terms of clinical important outcomes. For instance, if we are treating claudicants, as the majority of the patients treated with this devices, the adequate outcome should be quality of life measured by validated questionnaires. Amputation rate as an outcome in claudication is not a significant endpoint. So far, drug-eluting balloons in below-the-knee disease have shown no superiority over plain balloon angioplasty besides surrogate endpoints.

The 2017 ESC/ESVS Guidelines on the Diagnosis and Treatment of PADs don't attribute strong evidence to their use (class IIb recommendation) and recognize that there are some gaps in this field.[12]

To overcome the limitations of the sponsored trials, some independent trials are ongoing. The SWEDEPAD Trial is a multicenter, prospective Randomized Controlled Clinical Trial based on the Swedish Vascular Registry (SWEDVASC) Platform (governmental sponsor), with relevant clinical outcomes. It tests the hypothesis that drug eluting (DE) technology is superior to conventional endovascular treatment (no-DE) in terms of important clinical outcomes, when applied on infrainguinal (femoropopliteal and/or infrapopliteal) obstructive vascular lesions. The Study started by November 2014, estimates to enroll 3800 participants and the estimated study completion date is June 2021. The primary endpoint for patients with critical limb ischemia (SWEDEPAD 1 -NCT02051088) is amputation rate during follow-up and the primary endpoint for patients with intermittent claudication (SWEDEPAD 2 - NCT02051088) is health-related quality of life after one year.

Direct drug delivery

Direct drug delivery can be achieved via the *"bullfrog"* micro infusion catheter, currently, the only device in the market under the category of a direct drug delivery system. This micro infusion catheter comprises of a microneedle folded within a balloon. The device is deployed by expanding the balloon against the wall of the vessel and extending the needle into the vessel wall with penetration into the adventitial layer. Pharmacotherapies can be deployed in the vessel segment of interest, presently, dexamethasone, but in the future, other pharmaceutical agents, biologics, biosimilar, and even stem-cell therapies can be deployed in this fashion.[13]

The DANCE Trial (Dexamethasone to the Adventitia to Enhance Clinical Efficacy After Femoropopliteal Revascularization) is a single arm multicenter study designed to evaluate the safety and efficacy of the Bullfrog device in the treatment of femoropopliteal lesions ≤15 cm in length. 262 patients were enrolled with symptomatic PAD and received either PTA or atherectomy with dexamethasone infusion.[13] No major adverse events at 30 days and no device or drug related deaths at 12-months were reported, thus concluding safe use of the device in this vascular bed. At 12 months, the primary patency in the PTA + dexamethasone group and atherectomy + dexamethasone group were 75.5% and 78.4% respectively both of which exceeded the historical comparator of 72.3% determined by the authors. A limitation of this study is that it did not compare dexamethasone infusion to the other drug-eluting technologies.[13]

LIMBO trial (NCT02479555) applies the bullfrog device in lesions below the knee and has recently finished enrollment.

The advantages of direct drug delivery technology leads to an increased efficiency and precisions allowing for lower therapeutic concentrations of drugs to be used and smaller discreet segments (around 20mm each) to be treated. Since the therapy is delivered directly into the desired location, the risk of downstream washing of the agent is virtually non-existent. The micro-infusion catheter is a versatile device because the platform can be used to deliver any combination of drugs. However, the device is designed for use in treatment of heterogeneous and restenotic plaque but it is not suited for use in the treatment of homogenous and calcific plaque.

Covered stents

Covered stents are another strategy aimed to reduce restenosis, as it provides a mechanical barrier against tissue ingrowth. Nevertheless, edge stenosis limits this treatment option.[14] To overcome this problem, a combined technique

of preparation of the landing zones of the stent graft with DBC to prevent edge stenosis was proposed, although raising important cost issues.[15]

Bioresorbable stents

Another promising devices are the bioresorbable stents. The use of absorb everolimus-eluting stent used in infrapopliteal arteries showed promising three-years results, but further studies are warranted.[16] This was an industry sponsored trial with some limitations: single--center study, only 48 symptomatic patients (Rutherford category 3–6) with 27% of claudicants and treatment of infrapopliteal arteries, which is at least highly debatable. They treated small lesions ≤5 cm long (2 cm average). There were no control group, comparing results with other studies, but treating smaller lesions and different patients. Complete bioresorption occurs within 3 years, the results thereafter would be very newsworthy, yet in the meanwhile the results are similar to standard DES.[16]

Other

Many other strategies have been attempt both in animal studies and through innovative devices applied in humans.

There are agents abled to inhibit neointimal hyperplasia in animal models. Cinnamic aldehyde has anti--inflamatory properties, with a 61% reduction in vessel occlusion(17). The nontoxic red wine polyphenols loaded in a drug-eluting nanoparticle delivery system, highly specific for endothelial cells, showed to reduce smooth muscle cell proliferation and inflammatory cell and platelet activation, while promoting re-endothelization of the injured artery.[18] Honokiol is a natural bioactive product with anti tumor, anti inflammatory, anti oxidative, anti angio¬genic and neuroprotective properties.[19] This study determined that perivascular honokiol appli-cation reduced intimal thickening in rabbits 14 days after carotid artery injury, it may inhibit vascular smooth muscle cell (VSMCs) proliferation and reduce collagen deposition in local arteries.[19]

Owing to the rapidly dividing cells in the developing neointima, radiation therapy stands as an attractive approach to prevent intimal hyperplasia, especially when delivered locally via catheter as brachytherapy. The Randomized PARIS (Peripheral Arterial Radiation Investigational Study) failed to demonstrate a significant reduction in restenosis after PTA of femoropopliteal lesions.[20]

Combination of cold therapy with angioplasty (cryoplasty) is being considered for pilot tests in lower extremity arterial interventions on the basis of several reported small series and trials.[21] However, evidence that such therapy enhances efficacy and durability of angioplasty remains limited.[22]

Finally, Wang and co-authors generated a biomimetic delivery system using nanoclusters coated with platelet membranes to target the injured arterial wall. This technology makes the delivery system so specific for endothelial cells that it can be given intravenously.[23] These nanoclusters were loaded with an endothelium-protective epigenetic inhibitor (JQ1) or sirolimus and compared for their ability to mitigate restenosis without compromising the process of re-endothelization. JQ1/nanoclusters preserved the ability of the endothelium to recover while mitigating IH and appear to be a promising EC-protective candidate drug, suitable for next-generation anti-restenosis therapy.[23]

CONCLUSION

In conclusion, one might say that nowadays we are able to do great things, amazing procedures to our patients and it would be great if we could make them last. The complete understanding and control of intimal might be the answer.

REFERENCES

1. Kijani S, Vazquez AM, Levin M, Boren J, Fogelstrand P. Intimal hyperplasia induced by vascular intervention causes lipoprotein retention and accelerated atherosclerosis. Physiol Rep. 2017;5(14).
2. Kleinstreuer C, Hyun S, Buchanan JR, Jr., Longest PW, Archie JP, Jr., Truskey GA. Hemodynamic Parameters and Early Intimal Thickening in Branching Blood Vessels. Crit Rev Biomed Eng. 2017;45(1-6):319-82.
3. Haruguchi H, Teraoka S. Intimal hyperplasia and hemodynamic factors in arterial bypass and arteriovenous grafts: a review. J Artif Organs. 2003;6(4):227-35.
4. Franzoni M, Cattaneo I, Longaretti L, Figliuzzi M, Ene-Iordache B, Remuzzi A. Endothelial cell activation by hemodynamic shear stress derived from arteriovenous fistula for hemodialysis access. Am J Physiol Heart Circ Physiol. 2016;310(1):H49-59.
5. Dake MD, Ansel GM, Jaff MR, Ohki T, Saxon RR, Smouse HB, et al. Durable Clinical Effectiveness With Paclitaxel-Eluting Stents in the Femoropopliteal Artery: 5-Year Results of the Zilver PTX Randomized Trial. Circulation. 2016;133(15):1472-83; discussion 83.
6. Muller-Hulsbeck S, Keirse K, Zeller T, Schroe H, Diaz-Cartelle J. Long-Term Results from the MAJESTIC Trial of the Eluvia Paclitaxel-Eluting Stent for Femoropopliteal Treatment: 3-Year Follow-up. Cardiovasc Intervent Radiol. 2017;40(12):1832-8.
7. Tepe G, Laird J, Schneider P, Brodmann M, Krishnan P, Micari A, et al. Drug-coated balloon versus standard percutaneous transluminal angioplasty for the treatment of superficial femoral and popliteal peripheral artery disease: 12-month results from the IN.PACT SFA randomized trial. Circulation. 2015;131(5):495-502.
8. Thieme M, Von Bilderling P, Paetzel C, Karnabatidis D, Perez Delgado J, Lichtenberg M, et al. The 24-Month Results of the Lutonix Global SFA Registry: Worldwide Experience With Lutonix Drug-Coated Balloon. JACC Cardiovasc Interv. 2017;10(16):1682-90.
9. Schroe H, Holden AH, Goueffic Y, Jansen SJ, Peeters P, Keirse K, et al. Stellarex drug-coated balloon for treatment of femoropopliteal arterial disease-The ILLUMENATE Global Study:

12-Month results from a prospective, multicenter, single-arm study. Catheter Cardiovasc Interv. 2018;91(3):497-504.
10. Katsanos K, Spiliopoulos S, Kitrou P, Krokidis M, Karnabatidis D. Risk of Death Following Application of Paclitaxel-Coated Balloons and Stents in the Femoropopliteal Artery of the Leg: A Systematic Review and Meta-Analysis of Randomized Controlled Trials. J Am Heart Assoc. 2018;7(24):e011245.
11. Zeller T, Baumgartner I, Scheinert D, Brodmann M, Bosiers M, Micari A, et al. Drug-eluting balloon versus standard balloon angioplasty for infrapopliteal arterial revascularization in critical limb ischemia: 12-month results from the IN.PACT DEEP randomized trial. J Am Coll Cardiol. 2014;64(15):1568-76.
12. Aboyans V, Ricco JB, Bartelink MEL, Bjorck M, Brodmann M, Cohnert T, et al. Editor's Choice - 2017 ESC Guidelines on the Diagnosis and Treatment of Peripheral Arterial Diseases, in collaboration with the European Society for Vascular Surgery (ESVS). Eur J Vasc Endovasc Surg. 2018;55(3):305-68.
13. Razavi MK, Donohoe D, D'Agostino RB, Jr., Jaff MR, Adams G, Investigators D. Adventitial Drug Delivery of Dexamethasone to Improve Primary Patency in the Treatment of Superficial Femoral and Popliteal Artery Disease: 12-Month Results From the DANCE Clinical Trial. JACC Cardiovasc Interv. 2018;11(10):921-31.
14. Nishimoto Y, Fukuhara R, Miyamoto T, Toma M, Miyata A, Sato Y. Edge restenosis of a polytetrafluoroethylene-covered stent in the superficial femoral artery: insights from an imaging assessment with an electronic high-resolution angioscope. Cardiovasc Interv Ther. 2019.
15. Lin TC, Huang CY, Chen PL, Lee CY, Shih CC, Chen IM. Edge Stenosis After Covered Stenting for Long Superficial Femoral Artery Occlusive Disease: Risk Factor Analysis and Prevention With Drug-Coated Balloon Angioplasty. J Endovasc Ther. 2018;25(3):313-9.
16. Varcoe RL, Thomas SD, Lennox AF. Three-Year Results of the Absorb Everolimus-Eluting Bioresorbable Vascular Scaffold in Infrapopliteal Arteries. J Endovasc Ther. 2018;25(6):694-701.
17. Buglak NE, Jiang W, Bahnson ESM. Cinnamic aldehyde inhibits vascular smooth muscle cell proliferation and neointimal hyperplasia in Zucker Diabetic Fatty rats. Redox Biol. 2018;19:166-78.
18. Dugas TR, Brewer G, Longwell M, Fradella T, Braun J, Astete CE, et al. Nanoentrapped polyphenol coating for sustained drug release from a balloon catheter. J Biomed Mater Res B Appl Biomater. 2019;107(3):646-51.
19. Wang Y, Zhao D, Sheng J, Lu P. Local honokiol application inhibits intimal thickening in rabbits following carotid artery balloon injury. Mol Med Rep. 2018;17(1):1683-9.
20. Waksman R, Laird JR, Jurkovitz CT, Lansky AJ, Gerrits F, Kosinski AS, et al. Intravascular radiation therapy after balloon angioplasty of narrowed femoropopliteal arteries to prevent restenosis: results of the PARIS feasibility clinical trial. J Vasc Interv Radiol. 2001;12(8):915-21.
21. Wildgruber MG, Berger HJ. Cryoplasty for the prevention of arterial restenosis. Cardiovasc Intervent Radiol. 2008;31(6):1050-8.
22. McCaslin JE, Andras A, Stansby G. Cryoplasty for peripheral arterial disease. Cochrane Database Syst Rev. 2013(8):CD005507.
23. Wang B, Chen G, Urabe G, Xie R, Wang Y, Shi X, et al. A paradigm of endothelium-protective and stent-free anti-restenotic therapy using biomimetic nanoclusters. Biomaterials. 2018;178:293-301.

LUNG HERNIA RELATED WITH A ROPE BULLFIGHT: CASE REPORT

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Abstract

Lung herniation is an uncommon entity which was fully classified in 1845 after the study of several case reports. Acquired lung hernia, especially traumatic, is the most common etiology. In the absence of clear guidelines, management of lung hernia is made in a case-by-case basis. We present an asymptomatic middle lobe hernia perceptible on physical examination, but diagnosed initially by imaging studies. Patient medical history included a blunt bull trauma fourteen years before.

INTRODUCTION

Roland was the first to describe lung hernia (LH) in 1499, a rare condition defined as a protrusion of lung tissue through one of its bounding structures [1-10]. Despite being usually asymptomatic,[8,11] the diagnosis of LH is most frequently clinical[1,2,4,7,9] but it should be confirmed by imaging.[1,5] We present a case of an asymptomatic middle lobe hernia possibly after a blunt trauma.

CASE REPORT

A 73-year-old male was admitted to an intensive care unit for non-invasive ventilation with an exacerbation of COPD (chronic obstructive pulmonary disease). The thoracic computed-tomography (CT) showed herniation of the lateral segment of the middle lobe through the 6th intercostal space,with no signs of incarceration (Figures 1-3). The patient had a previous thoracic CT that confirmed the hernia presence for at least 3 years, although it was not perceptible in previous Chest x-Rays (CxR) (Figure 4).

When inquired, the patient recalled getting hitt and stepped on during a rope bullfight fourteen years before, resulting in multiple rib fractures.. His medical history included severe obesity, obstructive sleep apnea, , hypertension, diabetes, myocardial revascularization (3

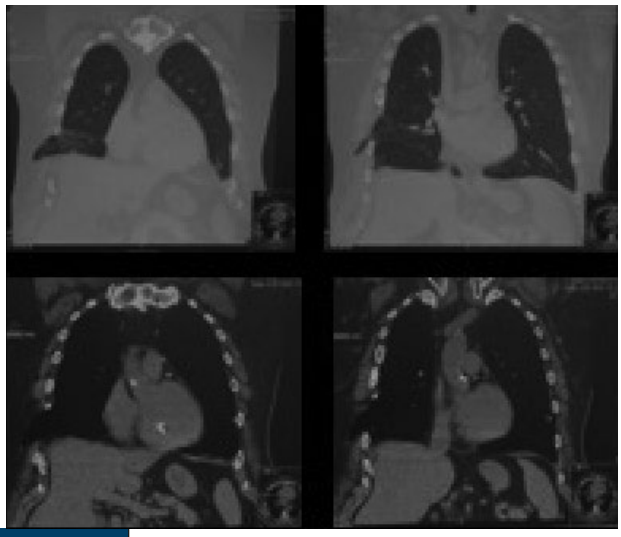

Figure 1 *Coronary CT views of middle lobe hernia.*

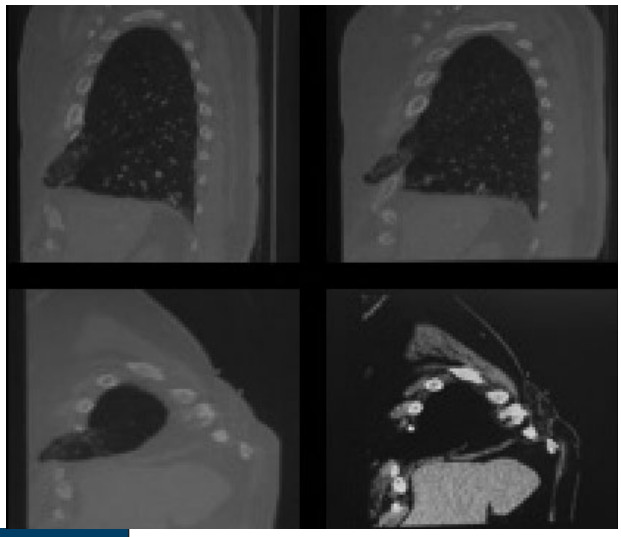

Figure 2 *Sagittal CT views of middle lobe hernia.*

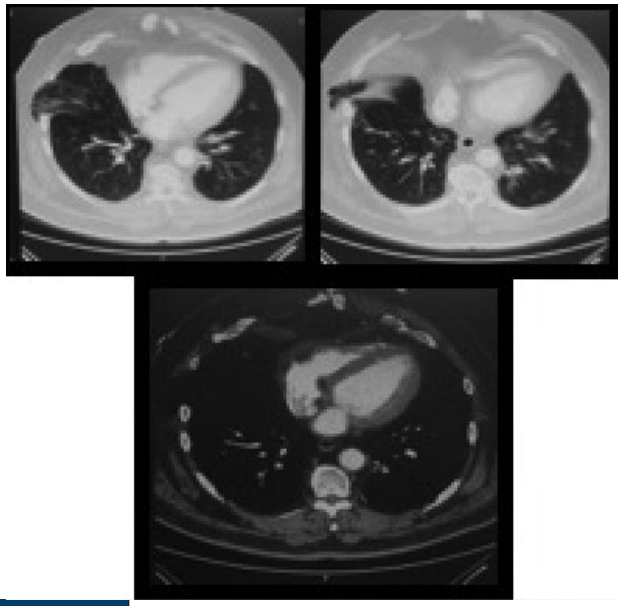

Figure 3 *Axial CT views of middle lobe hernia.*

years before) and ischemic heart failure (class II NYHA). The patient was on steroids for clinical exacerbations of COPD.

The patient noticed a mass on his right chest wall some years after the trauma, not perceptible with normal breathing. He denied hemoptysis, chest pain, subcutaneous emphysema, fever or any other symptoms. On physical examination there was a soft subcutaneous mass, which varied in size with breathing, bulging with cough and Valsava maneuvers (Figure 5).

The medical-surgical team agreed that being totally asymptomatic there was no indication for surgical treatment. The patient was discharged after resolution of the COPD's exacerbation.

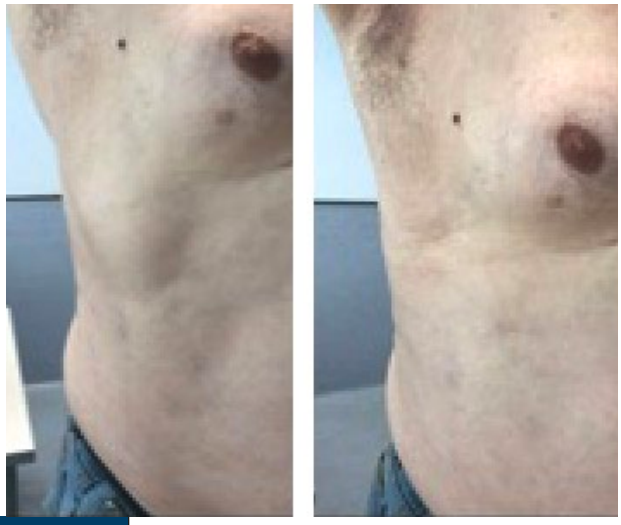

Figure 5 *Subcutaneous mass while performing Valsalva maneuvers.*

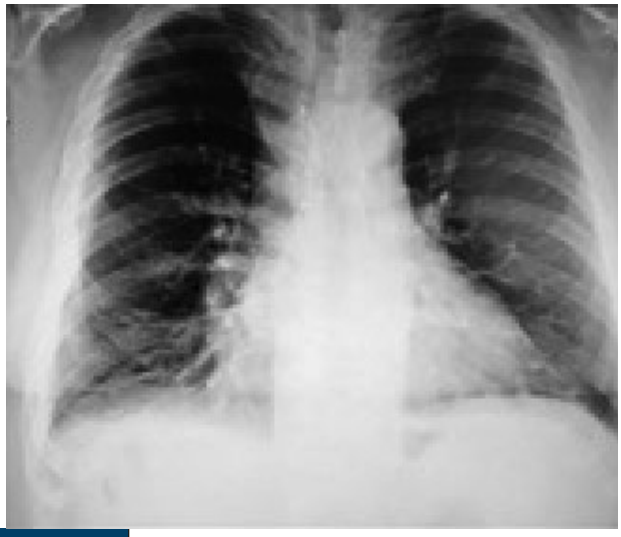

Figure 4 *Chest x-Ray on admission.*

DISCUSSION

In 1845, Morel-Lavallee classified LH etiologically and anatomically.[1-4,6, 8-9,12] Anatomically, it can be divided into cervical, intercostal, diaphragmatic or mediastinal.[2,3] Intercostal LH are the most common ones.[5,6,9,10] Lung herniation most frequently occurs anteriorly near the sternum or posteriorly near the vertebrae where there is only a single layer of intercostal muscles.[1,3,10,11] According to a database review on all patients who underwent pulmonary herniorraphy between 1991 and 2011, Seder et al. concluded that LH most frequently occur on the right side, fifth intercostal space, and is chronic in nature.[9]

Etiologically, LH is classified as congenital or acquired. Congenital hernias account for approximately 20% of the cases and are related to costal or cartilage malformations such as rib or intercostal hypoplasia or agenesis, or simply to an attenuation of the endothoracic fascia.[1,4, 8,10,11] They usually occur at the thoracic inlet or intercostal space, because of the absence of intercostal muscles and weakness of the fascia and can present only in adult life.[2,3] The supraclavicular region is the most frequent location of congenital LH.[5]

Acquired LH is the most common cause (80%).[1,2,4, 6,7,8-11] It can be classified as spontaneous, pathological, traumatic or post-surgical.[2-4,11] Predisposing factors are environmental and operative trauma, neoplastic or inflammatory processes, COPD and chronic steroid use.[3,4,9,10] Male sex, obesity and smoking are also described as risk factors.[9,10] Positive pressure ventilation and diabetes mellitus were also considered as potential risk factors.[8,9]

The mechanism of spontaneous LH involves an intercostal defect associated with increased intrathoracic pressure (intense or persistent coughing, sneezing, playing wind musical instrument, heavy weight lifting), in the presence of the risk factors previously described.[2-4,10] Castro et al. reported that spontaneous LH are mainly caused by Valsalva maneuvers.[6] They are typically more frequent in patients with COPD.[1]

Traumatic Lung Hernia (TLH) is an uncommon condition which may be benign or constitute an emergency.[4,6,11] It may appear immediately after the insult or be delayed for months or years.[1,2-4,6,8] They Usually occur on the anterior thoracic wall, where external intercostal muscles are absent and muscles are weaker.[1,4] Most of the times, TLH is due to the disruption of intercostal muscles or rib fractures, after trauma or after cardiopulmonary resuscitation; they can also occur after thoracic surgery.[3-7] Interestingly, postoperative LH is more common after minimally invasive procedures due to a less meticulous closure of the chest wall through the smaller skin incision.[2,3,5-7,8] Incidence of TLH is increasing related to an increment of high velocity penetrating weaponry.[14]

Pathological lung hernias are the least common variety.[1,3] They can be due to breast or chest wall pathology (malignant tumors, empyema necessitans, abcess, osteomyelitis or even tuberculosis), but its incidence decreased due to an early detection and treatment of these conditions.[1,3, 6,11]

According to the Morel-Lavallee classification, the case reported corresponds to an acquired thoracic pulmonary hernia with a traumatic origin. Nonetheless, it could also have been enforced by an exacerbation of the COPD. The patient also had other risk factors: poor tissue quality and healing capacity resulting from diabetes, obesity, smoking history and oral steroid use.

Physical findings are usualy similar to the present case.. Often, when there is no palpable mass it can be necessary to perform Valsalva maneuvers in order to detect the disorder.[1-4,11] Other presentation symptoms can be chest pain, dyspnea, subcutaneous emphysema, hemoptysis, bone crepitation, ecchymosis or chest wall instability.[1,3,4,6] In rare circumstances, diagnosis results from an accidental finding in imaging studies. A CxR especially during a Valsalva maneuver, can establish the diagnosis, but in the case of intercostal LH it could go undetected as in our case.[1-4,8-11] CT can confirm a herniated lung, the hernia sac and orifice in the chest wall as well as their anatomic relation with chest wall muscles.[3,4,8,9] It can also exclude complications such as incarceration.[3,8,10]

When it comes to treatment, a multidisciplinary team constituted by thoracic, general and sometimes a plastic surgeon, is recommended for decision making.[12] Classification is outdated, not guiding operative approaches or repair strategies in the modern era. Kuckelman et al. reinforce the idea that there is scant literature guiding managing of TLH with no evidence-based consensus.[14] It depends on the clinical condition of the patient but literature is ambivalent.[1,3,4,7,8,10] In most cases, LH needs no immediate repair or, if asymptomatic, any repair at all.[2,7,9,11] It is reasonable to wait for spontaneous regression before intervention, because complications are rare.[3,8,11] Some authors advocate conservative treatment like compression with pads and corsets, treating the underlying cause and promoting weight loss.[1,7] The majority of literature agree that large LH (or those with progressive increase in size) should be treated surgically, just like the incarcerated, strangulated or symptomatic ones, according to the patient conditions and surgical risk.[1-5,7,8-10,12] Associated lesions like rib fractures and chest wall defects should also be treated on a case-by-case basis.[6]

Repair techniques vary, ranging from primary closure to implantation of a mesh or muscle flaps.[7,9] In the majority of cases, primary closure of the defect produces good results, mainly for smaller defects.[1,7] Some authors advocate the use of prostetic mesh when the thoracic wall defect is large.[7,8] According to others, muscle flaps are usually preferable in this situation.[6] More recently Kuckelman et al. advocated the primary suture closure for defects less than 2 cm and a prosthatic mesh, if larger than 2 cm.[12]

In the presented case, LH was asymptomatic and without incarceration signs. So we've decided for a conservative approach, with a regular clinical and radiographic follow-up, which is mandatory.[8,9]

CONCLUSION

It is important to recognize LH as a benign disorder in most of the cases. Highly sensitive imaging such as CT, the increased prevalence of high-velocity blunt trauma and the increment of VATS (video-assisted thoracic surgery) worldwide, had increased the number of case reports.[9,12] There must be a meticulous technique when closing primary thoracotomies, mainly in COPD, obese, smoker, diabetic and chronic oral steroid user patients.

LH can be challenging to the thoracic surgeon. The classification system provides few orientations to clinical decision and operative repair and there is controversy regarding concomitant intraoperative decisions such as rib fixation and chest wall reconstruction.[6] Management depends on symptoms, location and size.[2,7] Indications for surgery are increasing size, pain or any signs of impending incarceration.[1,2,5,9,10] Repair for cosmetic reasons can be justified.[1,2,4,9,10] Approach must provide an optimal outcome in terms of recurrence, post-operative pain, hernia reduction and repair.

Because it is rare and variable, there are insufficient evidence-based studies to recommend a standard management of LH.

REFERENCES

1. Goverde P, Van Schill P, Van den Brande F, Vanmaele R. Chronic herniation of the lung in a patient with chronic obstructive pulmonary disease. Case report and review of the literature. Thorac Cardiovasc Surg 1998; 46 (3): 164-6.
2. Weissberg D. Lung hernia – A review. Adv Clin Exp Med 2013; 22 (5): 611–613.
3. Detorakis EE, Androulidakis E. Intercostal lung herniation – The role of imaging. J Radiol Case Rep 2014; 8(4): 16-24.
4. Marsico G, Boasquevisque C, Loureiro G, Merques R, Clemente A. Hérnia traumática do pulmão. Ver Col Bras Cir 2011; 38 (1): 77-8.

5. Ishida A, Oki M, Saka H, Seki Y. Postoperative intercostal lung hernia. Respirol Case Rep 2018; doi: 10.1002/rcr2.323
6. Castro PP, Machicao FV, Cohen RS, Silva RB, Rivera F. Minimally invasive management of traumatic lung herniation. J Surg Case Rep 2017; doi: 10.1093/jscr/rjx130.
7. Chiang TY, Yiu MF, Yang SM, Chau KC. Thoracoscopic management of incarcerated lung herniation after blunt chest trauma: a case report and literature review. J Thoracic Dis 2017; doi: 10.21037/jtd.2017.03.41
8. Puah SH, Tan Pei Sze C, Abisheganaden J. Respir Med Case Rep 2016; 20: 61-63.
9. Seder CW, Allen MS, Nichols FC, Wigle DA, Shen KR, Deschamps C, Cassivi SD. Primary and prosthetic repair of acquired chest Wall hernias: a 20-year experience. Ann Thorac Surg 2014; 98 (2): 484-9.
10. Corten BJ, Dijk WA, Bilderbeek-Beckers M, Janzing HM. Traumatic herniation of the lung. Acta Chir Belg 2016; 116(5): 322-324.
11. Sereide O, Stedjeberg JO. Traumatic intercostal pulmonary hernia. Injury 1975;7(1): 61-2.
12. Kuckelman J, Karmy-Jones R, Windell E, Izenberg S, David JS, Long W, Martin MJ. Traumatic thoracic rib cage hernias: Operative management and proposal for a new anatomic-based grading system. Am J Surg 2018; 215 (5): 794-800.

COLD AGGLUTININS AND CARDIAC SURGEY: A CASE REPORT

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Abstract

Cold agglutinins (CA) are autoantibodies whose clinical significance depends upon titer and thermal amplitude. Patients, which undergo cardio-pulmonary bypass and especially hypothermic cardioplegia myocardial protection, represent a challenge regarding operative management, as tissue temperature should be maintained above the threshold of agglutination. We report on a case in which the presence of CA was discovered during elective aortic valve replacement surgery, and managed with normothermic cardiopulmonary bypass and continuous retrograde warm blood cardioplegia administration.

INTRODUCTION

Cold agglutinins (CA) are IgM autoantibodies that react against I-antigens on the surface of erythrocytes. CA may be idiopathic, or more frequently, secondary to infective or neoplasic/lymphoproliferative disorders, among others.[1-5]

Systemic hypothermia and hypothermic cardioplegia delivery (usually 1-8ºC) is prone to induce CA related hemagglutination which can result in intracoronary thrombosis, incomplete cardioplegic delivery and high pressures within the circuit.

As the evidence regarding CA during cardiac surgery is mainly derived from case reports, management strategies remain controversial.

We report on a patient with accidental finding of cold agglutinin during elective aortic valve replacement surgery.

CASE REPORT

An 82-year-old female was admitted to our hospital with the diagnosis of severe aortic stenosis. Patient's anamnesis was unremarkable regarding frequent cold agglutinin disease associated findings: no history of hemolytic anemia, hemoglubinuria, acrocyanosis, livedo reticularis, previous infection or proliferative disorder was noted.

Anesthesia, cannulation and extracorporeal circulation were performed in the usual manner. An activated clotting time superior to 400 seconds was achieved with heparin (3mg/kg). The patient was cooled to a core temperature of 34ºC and myocardial protection was achieved with anterograde cold blood cardioplegia (Buckberg type with a 4:1 blood to BBRAUN™ crystalloid solution) after cross-clamping the aorta.

During administration of anterograde cardioplegia macroscopic precipitates, suggestive of agglutination, were visible within the cardioplegia circuit. This prompted immediate cessation of the anterograde cardioplegia administration; consultation of the imunohemotherapy service; and opening of the ascending aorta for further inspection. As agglutination was limited to the cardioplegia circuit, which was cooled to 4ºC, a temperature related agglutination phenomena was suspected and retrograde (via coronary sinus cathether) warm blood cardioplegia was initiated in order to wash-out any agglutination debris and provide further myocardial protection. Additionally, the patient was re-warmed and careful attention was paid to the temperature of the operating room and of the perfusions being administrated.

Aortic valve replacement was achieved with a St Jude Medical Trifecta™ GT™ 21-mm bioprosthesis (St. Jude Medical, Inc., St. Paul, MN, USA), performed with interrupted sutures in supra-annular position. The lowest core temperature registered was 34ºC. Intra-operative transesophageal echocardiography reveled no significant findings.

Immediate post-operative period was uneventful. No significant coagulopathy or chest drainage was noted. Blood transfusion and iron and eritrophoetin

supplementation were necessary due to mild hemolysis. The nadir of hemoglobin concentration was 7.4 g/dL. Maximum lactase dehydrogenase and bilirubin concentration were 375 U/L and 0,76 mg/dL respectively.

The patient was discharged from the hospital on postoperative day 12, with no evidence of active hemolysis.

Although the primary cause of CA in this patient was not made clear by the investigation that followed, the patient was ambulatory and clinically asymptomatic at 12 months follow-up.

DISCUSSION

Clinical significance of CA depends upon titer and the temperature below which antibody activation occurs. Low-titer CA (about 1:16) can be found in the sera of healthy individuals.[5] Evidence suggests that patients with low--titer and very low temperature reacting antibodies may undergo operation without changing management plan.

Patients with high-titer, high-temperature reacting CA undergoing routine CPB represent a challenge regarding operative management as tissue temperature should be maintained above the threshold of agglutination. Normothermic CBP with several myocardial protection techniques have been described, including intermittent cross-clamping, ventricular fibrillation, cold crystalloid cardioplegia and warm blood cardioplegia.[1-2,4-7]

Plasmapherisis and high dose immune globulin have been reported as effective methods of titer reduction. The former, however, is associated with large volume shifts, risk of infection and altered hemostasis and should be reserved for patients undergoing planned deep hypothermic arrest.[5-6]

The rationale regarding the choice of myocardial protection presented in this case rests in the evidence available in the literature. Patients with high titers CA represent a group in which warm cardioplegic myocardial protection may be indicated. Although still controversial, studies suggest that warm cardioplegia may be as effective as hypothermic cardioplegia regarding myocardial protection.[4]

Continuous cardioplegia infusion is beneficial in the sense that maintains tissue perfusion and prevents microvascular clotting by continuous washout.

Even though the peri-operative course of this patient was rather uneventful, adequate clinical follow-up and primary cause investigation are of utmost importance as may present prognostic implications.

CONCLUSION

CA are rarely of clinical importance, however, during cardiac surgery may represent origin of important complications. Pre-operative elaboration of a patient--oriented strategy is ideal to avoid CA related phenomena.

We report on a successful approach to CA using normothermic CPB and continuous warm blood cardioplegia.

REFERENCES

1. Kansaku R, Kuwaki K, Amano A, Inaba H, Tambara K, Yamamoto T, Sakakibara N. Aortic valve replacement to a patient with high titer of cold agglutinin. Ann Thorac Cardiovasc Surg. 2012;18(3):259-61.
2. Kalra A, Singh K, Sahoo M, Mahant TS. Cold agglutinin disease detected during open heart surgery. Indian J Hematol Blood Transfus. 2014 Mar;30(1):62-3.
3. Shah S, Gilliland H, Benson G. Agglutinins and cardiac surgery: a web based survey of cardiac anaesthetic practice; questions raised and possible solutions. Heart, Lung and Vessels. 2014; 6(3): 187-196.
4. Cho SH, Kim DH, Kwak YT. Normothermic cardiac surgery with warm blood cardioplegia in patient with cold agglutinins. Korean J Thorac Cardiovasc Surg. 2014 Apr;47(2):133-6.
5. Atkinson VP, Soeding P, Horne G, Tatoulis J. Cold agglutinins in cardiac surgery: management of myocardial protection and cardiopulmonary bypass. Ann Thorac Surg 2008; 85: 310-1.
6. Kanemitsu S, Onoda K, Yamamoto K, Shimpo H. Simple preoperative management for cold agglutinins before cardiac surgery. J Thorac Cardiovasc Surg. 2010 Nov;140(5):e73-4.
7. Barbara DW, Mauermann WJ, Neal JR, Abel MD, Schaff HV, Winters JL. Cold agglutinins in patients undergoing cardiac surgery requiring cardiopulmonary bypass. J Thorac Cardiovasc Surg. 2013 Sep;146(3):668-80.

CASOS CLÍNICOS CASE REPORTS

CIRURGIA NO ESCURO – UM CASO DE OCRONOSE CARDÍACA

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Resumo

A Alcaptonúria é uma doença genética rara, relacionada com o metabolismo da tirosina. As manifestações cardiovasculares são a forma de apresentação menos comum da doença, sendo a estenose aórtica a patologia mais frequentemente encontrada.

No presente artigo, apresentamos o caso de uma doente do sexo feminino de 72 anos proposta para cirurgia eletiva de substituição valvular aórtica com alterações intraoperatórias sugestivas de Ocronose Cardíaca.

Atendendo à raridade da doença, muito há por esclarecer acerca da sua história natural, nomeadamente no que se refere ao tipo de próteses utilizadas, motivo pelo qual é essencial a documentação e seguimento destes casos.

Abstract

Surgery in the dark – a case of Cardiac Ochronosis

Alkaptonuria is a rare genetic disorder related to tyrosine metabolism. The cardiovascular manifestations are rare being the aortic stenosis the most commonly reported.

We present a case of 72-year-old women who underwent aortic valve replacement with intraoperative findings in the aortic valve and the aortic wall suggestive of Cardiac Ochronosis.

Once it is a rare disease there are issues related to the natural history of the disorder that still unknown, namely the type of aortic prothesis in use.

For this reason, we find essential the documentation and follow-up of all these rare cases.

INTRODUÇÃO

A Alcaptonúria / Ocronose Alcaptonúrica é uma doença genética rara, de transmissão autossómica recessiva, relacionada com o metabolismo da tirosina.[1]

Tem uma incidência estimada que varia entre 1/250,000 a 1/ 1.000.000 de nascimentos.[2]

Deve-se à deficiência da oxidase do ácido homogentísico, a enzima responsável pela degradação metabólica da tirosina.[3]

A deficiência desta enzima resulta na excreção urinária de grandes quantidades de ácido homogentísico (alcaptonúria) e na acumulação do pigmento no tecido conjuntivo (ocronose), conferindo-lhes uma coloração mais escura.[3]

As manifestações clínicas mais frequentes da doença são então a coloração escura da urina (alcaptonúria); a pigmentação cutânea e ocular; e a artopatia.[3]

As manifestações cardiovasculares são menos comuns, sendo a estenose aórtica a alteração mais frequentemente encontrada.[4]

No presente artigo, apresentamos o caso de uma doente de 72 anos submetida a cirurgia de substituição valvular aórtica com achados intraoperatórios sugestivos de Ocronose Cardíaca.

CASO CLÍNICO

Doente do sexo feminino de 72 anos de idade, com história de coxartrose bilateral e cirurgia de prótese total do joelho; referenciada por estenose aórtica severa sintomática para tratamento cirúrgico.

O ecocardiograma pré-operatório revelou uma válvula aórtica tricúspide com com gradiente transvalvular

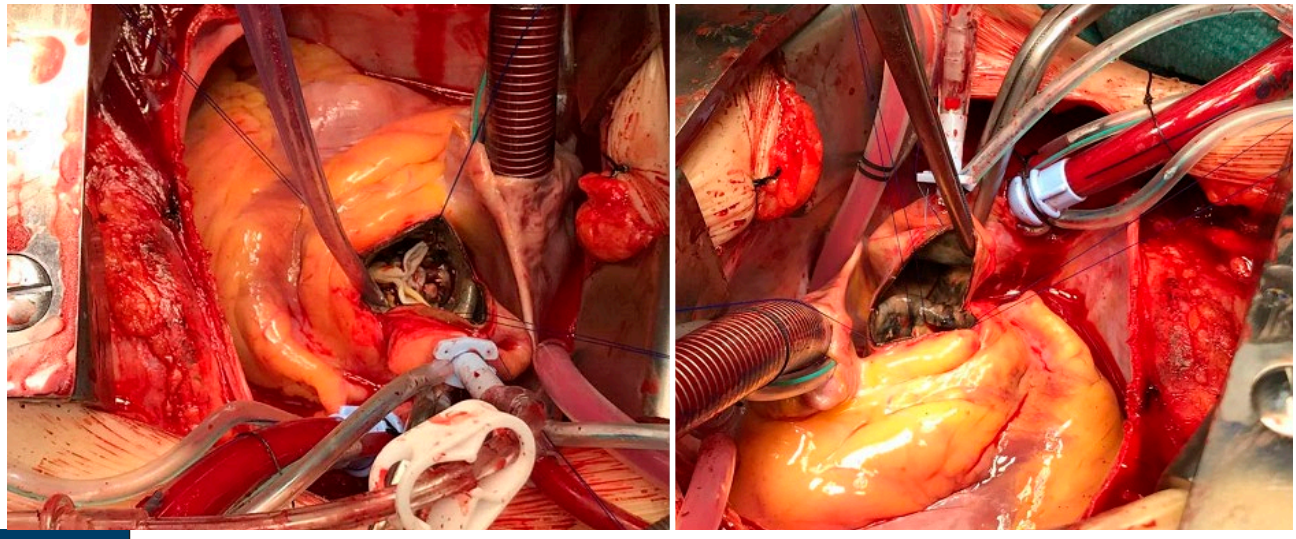

Figura 1 *Pigmentação escura da válvula aórtica e raiz da aorta*

máximo instantâneo de 82 mmHg e médio de 46 mmHg e área valvular aórtica de 0.8 cm2; além de hipertrofia concêntrica do ventrículo esquerdo e boa função sistólica global e segmentar bi-ventricular. A coronariografia era normal.

Intra-operatoriamente, observou-se uma válvula aórtica tricúspide com alterações degenerativas e com exuberante pigmentação escura dos seus folhetos. A coloração escura estendia-se à íntima da aorta ascendente e da raíz da aorta (Figuras 1 e 2).

Foi realizada substituição valvular aórtica por prótese biológica SOLO SMART nº 21, sem intercorrências.

O exame histopatológico da válvula aórtica removida, mostrou evidência de degenerescência mixóide e pigmentação acastanhada, com reação negativa após coloração histoquímica de Perls (ausência de ferro), sendo estes achados compatíveis com o diagnóstico de Ocronose (Figuras 3).

No exame físico da doente, havia evidência de pigmentação escura da esclerótica de ambos os olhos e dos

Figura 2 *Pigmentação escura dos folhetos da válvula aórtica removida (Macroscopia)*

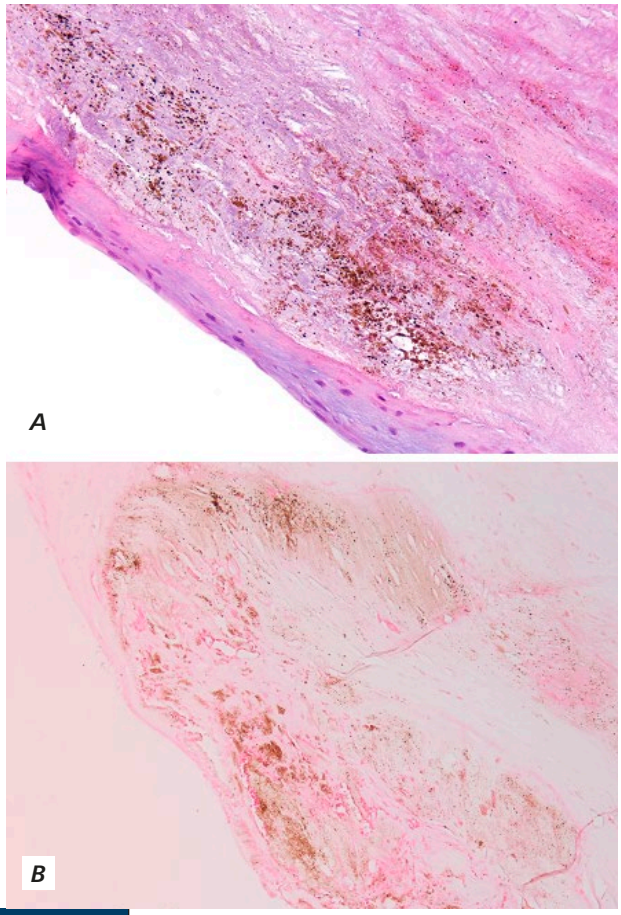

Figura 3 ***A**) HE 200x Maior detalhe do pigmento acastanhado, em àrea de degenerescência mixoide; **B**) Coloração histoquímica de Perls, pigmento negativo, mantendo-se acastanhado. (Ausência de Ferro)*

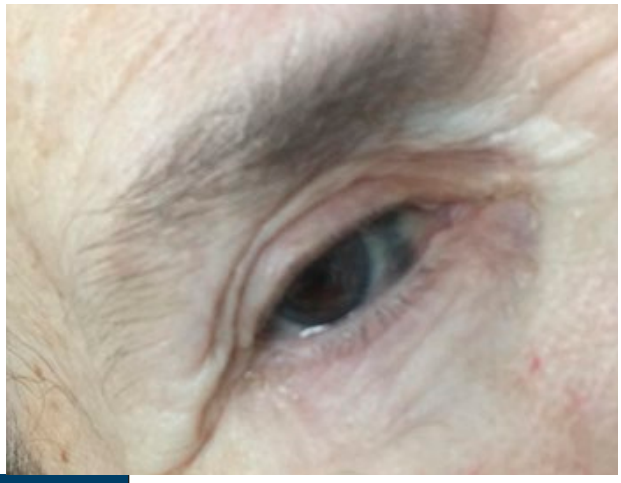

Figura 4 *Pigmentação escura da esclerótica.*

pavilhões auriculares (Figura 4). É também de referir na história prévia da doente, a presença de depósitos de pigmento escuro nos tecidos moles aquando da cirurgia de prótese total do joelho, além de coloração escura da urina (alcaptonúria); achados que suportam o diagnóstico.

A doente teve uma evolução favorável no pós-operatório, sem intercorrências para o tipo de intervenção realizada.

DISCUSSÃO

O envolvimento cardiovascular na Alcaptonúria/Ocronose Alcaptonúrica é raro, estando relacionado com a deposição de pigmento ocronótico nas válvulas cardíacas, artérias coronárias, parede da aorta, endocárdio e pericárdio.[5]

A extensa deposição extracelular de pigmento nas cúspides e anel aórtico leva a uma reação inflamatória com aceleração da calcificação distrófica e fibrose destas estruturas.[6,7]

A estenose valvular aórtica é aceite como a manifestação cardíaca mais frequentemente encontrada em pacientes com Ocronose.[4]

A incidência de doença valvular aórtica na quinta década de vida parece ser superior nos doentes com Ocronose Alcaptonúrica.[3,8]

Ainda assim, a maioria dos casos de estenose aórtica associados à Ocronose Alcaptonúrica documentados ocorreram na sexta e sétima décadas de vida, o que é sobreponível à população em geral.

Até ao momento, não existe um tratamento eficaz para a alcaptonúria, sendo a abordagem médica dirigida aos sintomas ou às complicações da doença. A vasta maioria dos doentes necessita de procedimentos ortopédicos ao longo da sua vida.[3]

No que respeita à Ocronose Cardíaca, mais especificamente à estenose aórtica associada à alcaptonúria, o tratamento passa pela substituição valvular aórtica, não existindo consenso em relação à escolha da prótese ideal.[9,10]

Atendendo à raridade da patologia, não se conhece o efeito dos depósitos de ácido homogentísico nas próteses biológicas.[9]

Por esse motivo, a escolha do tipo de prótese no doente apresentado teve como base as recomendações para a população geral, com especial atenção à possibilidade de futuras intervenções ortopédicas.

CONCLUSÃO

A Ocronose Cardíaca é uma patologia rara, muitas vezes diagnosticada aquando de uma cirurgia valvular aórtica eletiva, pelo que é essencial o reconhecimento das suas manifestações clínicas.

O follow-up dos doentes com alcaptonúria portadores de próteses valvulares, biológicas ou mecânicas, permitirá uma melhor compreensão da evolução da doença, o que se traduzirá numa melhor abordagem deste grupo de doentes.

REFERÊNCIAS

1. Janocha S, Wolz W, Srsen S, et al. The human gene for alkaptonuria (AKU) maps to chromosome 3q. Genomics. 1994;19:5-8.
2. La Du BN. Alkaptonuria. In: Scriver CR, Beandet AL, Sly WS, Valle D, eds. Metabolic & Molecular Basis of Inherited Diseases. 8th ed. New York, NY: McGraw-Hill; 2001: 2109-2123.
3. Phornphutkul C, Introne WJ, Perry MB, Bernardini I, Murphey MD, Fitzpatrick DL, et al. Natural history of alkaptonuria. N Engl J Med 2002;347(26):2111-21.
4. Steger, C. M. 2011. Aortic valve ochronosis: a rare manifestation of alkaptonuria. BMJ Case Rep. pii: bcr0420114119.
5. Magdy M El-Sayed Ahmed, MD, MSc, Omar Hussain, MD, MRCS, David A Ott, MD, FACS, FACC, and Muhammad Aftab, MD, FACC. Severe Aortic Valve Stenosis Due to Alkaptonuric Ochronosis. Seminars in Cardiothoracic and Vascular Anesthesia. Vol 21, Issue 4, pp. 364 – 366.
6. Gaines JJ Jr. The pathology of alkaptonuric ochronosis. Hum Pathol. 1989;20:40-46.
7. Helliwell TR, Gallagher JA, Ranganath L. Alkaptonuria: a review of surgical and autopsy pathology. Histopathology. 2008;53:503-512.
8. Gaines JJ Jr, Pai GM. Cardiovascular ochronosis. Arch Pathol Lab Med. 1987;111:991-994.
9. Selvakumar D, Sian K, Sugito S, Singh T. Ochronosis of the aortic valve. J Thorac Dis 2018;10(5):E332- E334. doi: 10.21037/jtd.2018.05.16
10. de Cima S., Rodriguez-Esteban A., Mencía P., Pilar, Junceda S., Albaiceta G. Un caso raro de valvulopatía causada por alcaptonuria. Cir Cardiov. 2018;25(1):38–40

HEMODIALYSIS ACESS – A CREATIVE ATTITUDE IS NECESSARY

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Abstract

Creating and maintaining a functional vascular access (VA) is a critical factor in the survival of a dialysis patient. It implies a creative attitude either to maintain its functionality or to build a new one wherever possible, being it autologous or synthetic.

We describe the VA history of a 59 years-old male patient, with extreme obesity, which started in 2012 with failed attempts of VA construction in both forearms until a functional brachiocephalic arteriovenous fistula (AVF) in the right upper limb was achieved. However, it required ligation due to severe venous hypertension secondary to central venous disease related to previous CVC use. As he had no good superficial conduit in the left arm we decided to harvest the arterialized right cephalic vein and implant it in the left arm, creating an autologous arteriovenous shunt between the brachial artery and axillary vein (AV). Despite initial patency, it failed irreversibly approximately one year after creation. As no more superficial veins were available in the upper limbs, a prosthetic access was the next step. We decided for a hybrid graft (HG) between the left brachial artery and the AV because of the patient's biotype and scarred axilla that impeded a safe re-intervention on the AV. This graft was used between 2015 and 2017 with multiple interventions to maintain patency. In 2017 a significant diffuse prosthesis deterioration and reduced AVF flow were noticed with no possible segmental reconstruction. We were then forced to proceed with subtotal graft substitution preserving the outflow stented segment of the HG, using an early cannulation graft to prevent CVC use. After this successful reconstruction, the patient started hemodialysis on the following day with no intercurrences registered.

INTRODUCTION

A detailed pre-operative history and physical examination is essential in the evaluation of a patient with end stage kidney disease and indication for a vascular access construction. Complementary venous and arterial ultrasound mapping provides crucial information for planning the access construction and decreases primary failure rate.[1-4]

After construction, surveillance is important to attest the vascular access function and patency and identify correctable lesions that may lead to access loss and it can be done during or outside the hemodialysis sessions.[5,6] Duplex ultrasound should also be the first line imaging method in patients with suspected dysfunction.[7,8]

However, even when the monitoring is correctly done, under certain circumstances the access failures to mature, develops dysfunction or thromboses which make us search for alternatives. However, we might be faced with exhausted venous conduits for the creation of a new access, requiring our creativity.

CLINICAL CASE

The authors describe the vascular access history of a 59-year-old male patient, with morbid obesity. The patient weighted around 130 kilograms with particularly central obesity and end stage kidney disease with requirement to enter a hemodialysis program.

His chronic kidney disease was secondary to diabetic nephropathy and his first vascular access was built in 2012. Planning the access creation was made with physical examination and ultrasound evaluation with venous and arterial mapping, once he achieved stage four of chronic renal disease, approximately six months before planned vascular access requirement.

Despite what we consider to have been a good planning and surgical construction, we had to deal with primary failure of several autologous accesses constructed in both upper limbs, not only bilateral radio-cephalic fistulas but also a left brachio-cephalic fistula, until it was possible to achieve the maturation of a brachio - cephalic fistula in the right upper limb. During this period, the patient progressed to five stage of chronic renal disease

and was inevitable to use a central venous catheter (CVC) in both internal jugular veins and the right subclavian vein. The thrombophilia study conducted was negative.

In 2013, the patient developed severe right upper limb venous hypertension, secondary to central venous stenosis, requiring fistula ligation. It was a normal debit fistula with 740ml/min, measured with Doppler ultrasound on the brachial artery, with turbulent flow and prolonged hemostasis time associated to venous hypertension. Angiography identified a long pre-occlusive central vein stenosis encompassing the right subclavian vein. Simple angioplasty was conducted at first but with restenosis before three months after the procedure. Due to the stenosis location and risk of eventual stent thrombosis, we performed a translocation of this cephalic vein and constructed a brachio-axillary vascular access in the left upper limb (Fig. 1). Meanwhile, it was necessary to use a new CVC in the femoral vein.

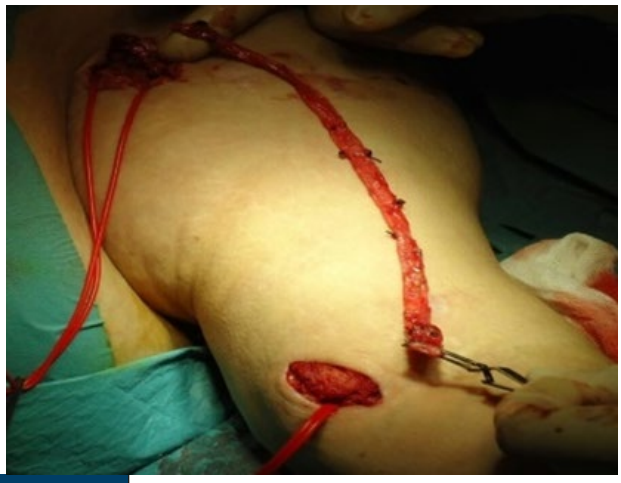

Figure 1 *Right cephalic vein translocation to the upper left limb.*

In 2014, one year after the cephalic translocation, the quality of dialysis through this access started to decrease significantly precipitated by a cephalic arch vein stenosis and aneurysmal degeneration that culminated with access thrombosis, despite simple angioplasties and stenting of the cephalic arch. On physical and ultrasound evaluation we identified a fusiform, with long extension, aneurysm with organized thrombus, which made us abandon this cephalic vein for further investment.

After aneurysmatic cephalic vein excision, we created a new brachio – axillary access with an hybrid graft. An open surgical approach to the brachial artery was used to perform the proximal anastomosis. Distal anchoring of the Gore Hybrid Vascular Graft on the axillary vein was delivered through a 14French introducer using the wire and "peel–away" technique (Fig. 2).

Between June 2014 and May 2015, despite several interventions with simple angioplasties due to juxta-anastomotic area stenosis to maintain the primary assisted patency of this hybrid Graft, the access thrombosed. Correction was made with surgical thrombectomy and creation of a new proximal anastomosis. Since May until October, simple angioplasties due to neo-intimal prosthetic hyperplasia were still performed.

During this period, difficult cannulation access was identified by the nurses at the hemodialysis center so as deterioration of the graft in the context of multiple punctures, despite resort to ultrasound assisted cannulation. That was the motive why we decided to extract an impaired graft segment and conduct an interposition graft with a ePTFE tubular graft (Fig.3).

Along the following period until 2017, the dialysis quality progressively decreased so as graft flow, reaching 350ml/min due to diffuse neointimal hyperplasia within the graft that caused hemodynamic effect with a decrease in lumen area superior to 70% . As a result, it was necessary to replace the graft. Since the distal segment anchored

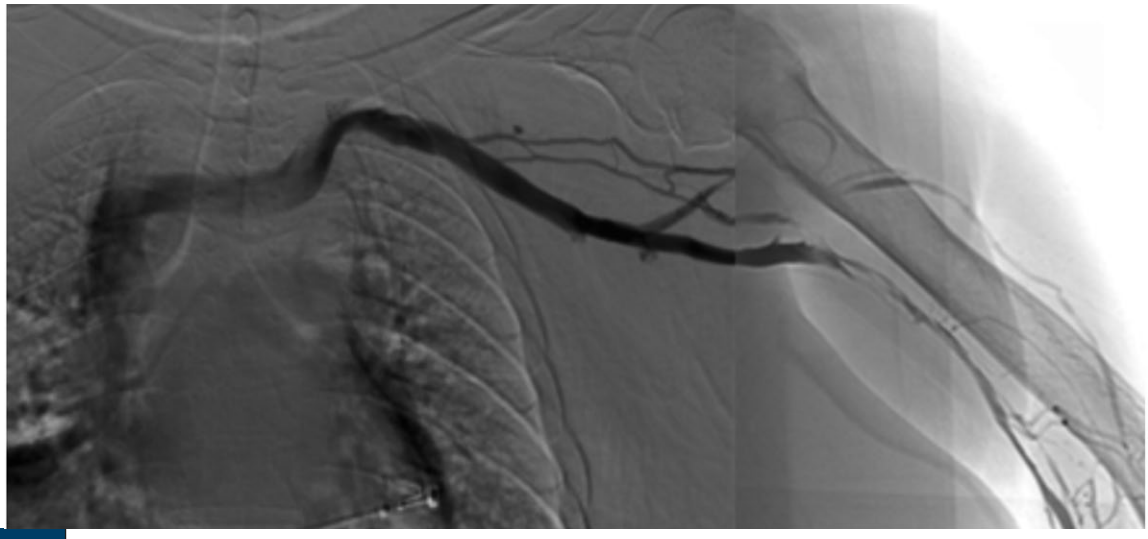

Figure 2 *Angiographic image after the Hybrid graft implantation.*

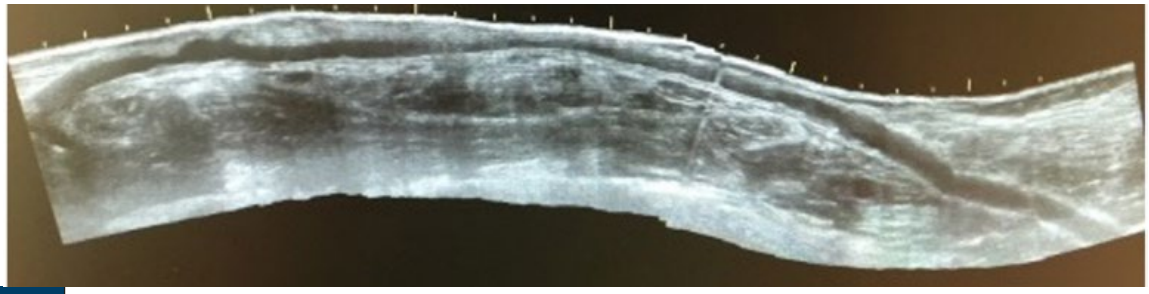

Figure 3 *Ultrasound image after the segmental interposition with a simple ePTFE Graft.*

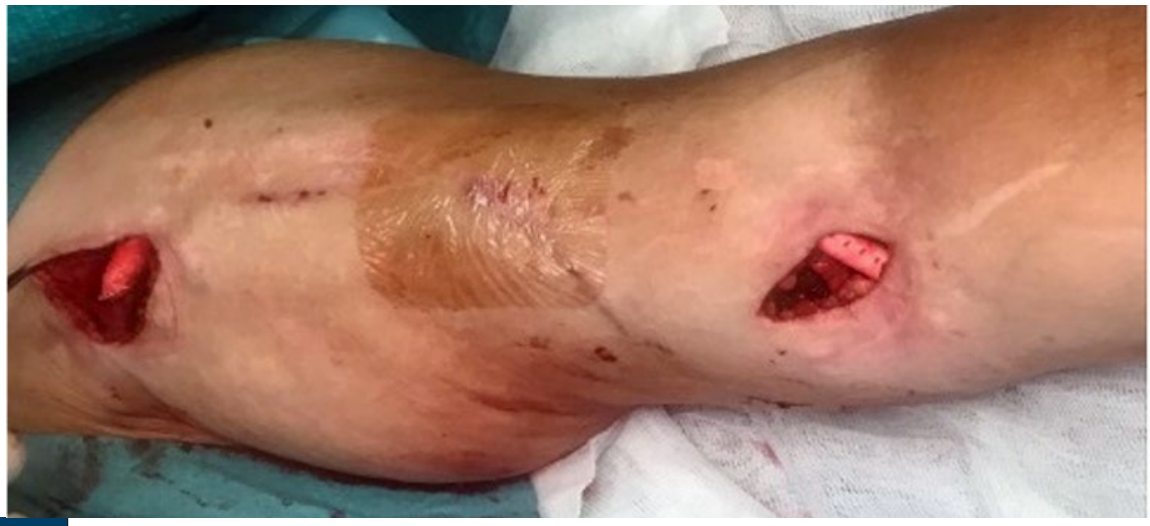

Figure 4 *Post implantation of the Early cannulation Graft.*

to the axillary vein was free of significant stenosis, it was possible to preserve that segment constructing an interposition graft with an early cannulation ePTFE graft (Gore Acuseal®) (Fig. 4).

This latter procedure was uneventful. There was no necessity of CVC use and the patient started hemodialysis, through this vascular access on the next day.

He has now 16 months of follow up and required only one simple angioplasty, eleven months after the procedure, due to distal juxta-anastomotic stenosis in order to preserve its primary assisted patency.

DISCUSSION / CONCLUSION

In general, central venous catheters are associated with inferior hemodialysis quality and reduced average life expectancy for this patients9. Therefore, all efforts should go towards building a functioning arteriovenous vascular access.

The appropriate vascular access for a patient depends on his individual available vessels and surgeon experience. Once this objective is reached, regular monitoring of vascular access and a dedicated team to treat any complications that may arise is necessary to achieve the best functionality possible.

In complex situations like the one here exposed, the vascular surgeon experience is essential, as well the capacity to promptly diagnose access failure in order to explore the residual vascular patrimony of the patient and to select the most adequate solution, whether by open or endovascular surgery. There are some published studies concerning the functionality of vascular accesses in obese patients as described by Micah R Chan et al and Kats M et al,[10,11] but the results are still debatable.

Considering the evolution of this particular vascular access so as the most recent published literature, once the functioning upper limb access was threatened by central venous stenosis, endovascular solutions should be considered. Still, there is some controversy about the best approach with some authors publishing good results, yet others presenting reduced primary patency on long-term follow up.[12,12] In our daily practice, we tend to favor endovascular treatment of this central vein stenosis but, in this particular case, once presented with early restenosis and considering the lesion characteristics so as absence of immediately accessible venous patrimony in the left upper limb for an autologous access construction and the existence of an arterialized and matured right cephalic vein made us lead to that solution, which is someway also described in the literature. Great saphenous vein and femoral vein translocation to the upper arm are commonly believed to have higher

complications rates, with poor maturation of the first one even considering good patency with the last one.[14-18]

The cephalic arch is prone to the development of hemodynamic significant stenosis being the most frequent location for stenosis of the upper arm dysfunctional vascular accesses, corresponding to 30-55% of all stenosis sites.[19] While endovascular solutions seem a reasonable option for the first pathology preventing an open surgical revision that might jeopardize the creation of a future basilic vein fistula, whether to choose between simple angioplasty, bare metal stent, graft stent or even drug eluting balloon is debatable with high recurrence rates.[20-22] Vascular access aneurysms are frequently accompanied by pre or post-aneurysm stenosis. In our case, the thrombosed aneurysm was complicated by wall-adherent thrombus which implied ligation of the aneurysmal section and graft interposition. The option for the hybrid graft was made considering the biotype of the patient and despite having considered basilic vein employment, the low basilic vein diameter (<6mm, both arms), an axillary zone with a previous surgical approach and necessity of continuous dialysis sessions, preferably without novel use of CVC, made us elect the construction of a new access with an hybrid prosthesis.

The future of prosthetic grafts for AV access appears bright as new approaches and technology are being investigated. Modifications in already existing grafts or development of new graft materials are being developed in an attempt to improve primary and secondary patency rates and reduce complications. Some grafts have also been upgraded with a silicone layer within the PTFE to allow early cannulation and to reduce needle hole bleeding.[23]

In this clinical case the use of a Hybrid Graft followed by simple graft and then early cannulation graft interpositions was described. Hybrid Grafts have hemodynamic advantages and also the possibility of anchoring its distal zone on what is expected to be a difficult surgical access region. Limitations come normally from the size of the graft that is 6 mm (according to the currently commercially available grafts) and of the outflow veins that should be larger than 4 mm. Other disadvantages are related to the published prosthesis infection rates that are around 7% and make this, together with the degradation at the puncture site, a long-term concernOn the other hand, early cannulation grafts allow an almost immediate cannulation and hemodialysis preventing unnecessary placement of a central venous catheter. The prosthesis is inserted into the vein, and the suture line is created on the outside surface of the prosthesis and is not located within the bloodstream at the graft-vein juncture with hemodynamic improvements. Nevertheless, prosthetic grafts still have significantly higher thrombotic threshold velocities than autologous grafts and require a higher flow to maintain patency.[24-32]

Acknowledgements: *To the Vascular Surgery Department of Centro Hospitalar Universitário do Porto*

Differential diagnosis should include malignancy and a special attention to this pathology in the transplant--recipient population should be present.

Despite mucormycosis being a rare infection, its prevalence is expected to raise together with increasing number and survival of the organ transplantation population as well as acquired immunodeficiencies. A high level of suspicion is recommended in the presence of the right clinical setting, as early diagnosis may be determinant for the prognosis.

REFERENCES

1. Clinical practice guidelines for vascular access. Am J Kidney Dis 2006;48:S176e247.
2. Beathard GA. Physical examination: The forgotten tool. In: Gray RJ, Sands JJ, editors. Dialysis access: a multidisciplinary approach. Philadelphia, PA: Lippincott Williams & Wilkins; 2002. p. 111e8. 381;
3. Centers for Medicare & Medicaid Services. 2004 Annual Report: ESRD clinical performance measures project. Baltimore, MD, Department of Health and Human Services, Centres for Medicare and Medicaid Services, Center for Beneficiary Choices, 2004. Am J Kidney Dis. 2005;46(Suppl 2):1. Available from: http://www.ajkd.org/article/S0272-6386(05)00922-4/pdf. Accessed Sep 12, 2017. 8.
4. Silva MB Jr, Hobson RW 2nd, Pappas PJ, et al: A strategy for increasing use of autogenous hemodialysis access procedures: Impact of preoperative noninvasive evaluation. J Vasc Surg 27:302-307, 1998
5. Trerotola SO, Scheel Jr PJ, Powe NR, Prescott C, Feeley N, He J, et al. Screening for dialysis access graft malfunction: comparison of physical examination with US. J Vasc Intervent Radiol JVIR 1996;7:15e20. 382 Schuman E, Ronfeld A, Barcl;
6. Robbin ML, Chamberlain NE, Lockhart ME, Gallichio MH, Young CJ, Deierhoi MH, et al. Hemodialysis arteriovenous fistula maturity: US evaluation. Radiology 2002;225:59e64
7. Mihmanli I, Besirli K, Kurugoglu S, Atakir K, Haider S, Ogut G, et al. Cephalic vein and hemodialysis fistula: surgeon's observation versus color Doppler ultrasonographic findings. J Ultrasound Med 2001;20:217e22;
8. Beathard GA. The treatment of vascular access graft dysfunction: a nephrologist's view and experience. Adv Ren Replace Ther 1994;1:131e47. 380;
9. Vascular Access 2006Work Group. Clinical practice guidelines for vascular access. Am J Kidney Dis 2006;48(Suppl 1):S248-73
10. Chan MR, Young HN, Becker YT, Yevzlin AS; Obesity as a predictor of vascular access outcomes: analysis of the USRDS DMMS Wave II study; Semin Dial. 2008 May-Jun;21(3):274-9. doi: 10.1111/j.1525-139X.2008.00434.x. Epub 2008 Apr
11. Kats M, Hawxby AM, Barker J, Allon M; Impact of obesity on arteriovenous fistula outcomes in dialysis patients; Kidney Int. 2007 Jan;71(1):39-43. Epub 2006 Sep 27
12. Yang HT et al; A prospective randomized study of stent graft placement after balloon angioplasty versus balloon angioplasty alone for the treatment of hemodialysis patients with prosthetic graft outflow stenosis; J Vasc Surg 2018 Aug;68(2):546-553. doi: 10.1016/j.jvs.2017.12.062. Epub 2018 Apr 2
13. Rangel LE; Primary Stenting Is Not Necessary in Benign

Central Venous Stenosis; Ann Vasc Surg 2018 Jan; 46:322-330. doi: 10.1016/j.avsg.2017.07.001. Epub 2017 Aug 12
14. Teruya TH, Schaeffer D, Abou-Zamzam AM, Bianchi C. Arteriovenous graft with outflow in the proximal axillary vein. Ann Vasc Surg 2009;23:95e8;
15. Rivers SP, Scher LA, Sheehan E, et al: Basilic vein transposition: An underutilized autologous alternative to prosthetic dialysis angioaccess. J Vasc Surg 18:391-396, 1993
16. Scott JD, Cull DL, Kalbaugh CA: The mid-thigh loop arteriovenous graft: Patient selection, technique and results. Am Surg 72:825-828, 2006 33.
17. Brownie ER, Kensinger CD, Feurer ID, Moore DE, Shaffer D: A comparison of patency and interventions in thigh versus Hemodialysis Reliable Outflow grafts for chronic hemodialysis vascular access. J Vasc Surg 64:1392-1399, 2016
18. Takuya Hatakeyama et al; Introduction of arteriovenous grafts with graft insertion anastomosis for hemodialysis access; J Vasc Surg 2017 Sep;66(3):952-957. doi: 10.1016/j.jvs.2017.03.441. Epub 2017 Jun 12
19. Rajan DK, Clark TWI, Patel NK, Stavropoulos SW, Simons ME. Prevalence and treatment of cephalic arch stenosis in dysfunctional autogenous hemodialysis fistulas. J Vasc Intervent Radiol JVIR 2003;14:567e73 (12761309);
20. Patanè D, Giuffrida S, Morale W, L'Anfusa G, Pulliatti D, Bisceglie P, et al. Drug-eluting balloon for the treatment of failing hemodialytic radiocephalic arteriovenous fistulas: our experience in the treatment of juxta-anastomotic stenoses. J Vasc Access 2014;15:338e43;
21. Portugaller RH, Kalmar PI, Deutschmann H. The eternal tale of dialysis access vessels and restenosis: are drug-eluting balloons the solution? J Vasc Access 2014;15:439e47;
22. Liu YH, Hung YN, Hsieh HC, Ko PJ. Surgical thrombectomy for thrombosed dialysis grafts: comparison of adjunctive treatments. World J Surg 2008;32:241-5.
23. Scher LA, Katzman HE: Alternative graft materials for hemodialysis access. Semin Vasc Surg 17:19-24, 2004 27;
24. Tordoir J, Canaud B, Haage P, Konner K, Basci A, Fouque D, et al. EBPG on vascular access. Nephrol Dial Transplant 2007;22:ii88e117;
25. Hazinedaroglu SM, Tuzuner A, Ayli D, Demirer S, Duman N, Yerdel MA. Femoral vein transposition versus femoral loop grafts for hemodialysis: a prospective evaluation. Transplant Proc 2004;36:65e7;
26. Huber TS, Hirneise CM, Lee WA, Flynn TC, Seeger JM. Outcome after autogenous brachial-axillary translocated superficial femoropopliteal vein hemodialysis access. J Vasc Surg 2004;40:311e8;
27. Kitrou PM, Katsanos K, Spiliopoulos S, Karnabatidis D, Siablis D. Drug-eluting versus plain balloon angioplasty for the treatment of failing dialysis access: final results and cost-effectiveness analysis from a prospective randomized controlled trial (NCT01174472). Eur J Radiol 2015;84:418e23;
28. Glickman M; Drug eluting grafts for hemodialysis access; J Vasc Access. 2017 Mar 6;18(Suppl. 1):53-55. doi: 10.5301/jva.5000671. Epub 2017 Mar 5
29. Berard X, Ottaviani N, Brizzi V, et al: Use of the Flixene vascular access graft as an early cannulation solution. J Vasc Surg 62:128-134, 2015 28;
30. Glickman MH, Burgess J, Cull D, et al: Prospective multicenter study with a 1-year analysis of a new vascular graft used for early cannulation in patients undergoing hemodialysis. J Vasc Surg 62:434-441, 2015;
31. Glickman MH, Burgess J, Cull D, Roy-Chaudhury P, Schanzer H; Prospective multicenter study with a 1-year analysis of a new vascular graft used for early cannulation in patients undergoing hemodialysis; J Vasc Surg. 2015 Aug;62(2):434-41. doi: 10.1016/j.jvs.2015.03.020. Epub 2015 May 4
32. Al Shakarchi J, Houston G, Inston N; Early cannulation grafts for haemodialysis: a systematic review; J Vasc Access. 2015 Nov-Dec;16(6):493-7. doi: 10.5301/jva.5000412. Epub 2015 Jun 12
33. Maytham GG, Sran HK, Chemla ES; The use of the early cannulation prosthetic graft (Acuseal™) for angioaccess for haemodialysis; J Vasc Access. 2015 Nov-Dec;16(6):467-71. doi: 10.5301/jva.5000390. Epub 2015 May 23
34. Windus DW, Jendrisak MD, Delmez JA. Prosthetic fistula survival and complications in hemodialysis patients: effects of diabetes and age. Am J Kidney Dis. 1992;19(5):448-452

SYNOVIAL HAEMANGIOMA OF THE KNEE – DIAGNOSIS BY MAGNETIC RESONANCE IMAGING

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Resumo

O hemangioma sinovial é uma rara malformação vascular não neoplásica da membrana sinovial descrita por Bouchut em 1856. Menos de 200 casos foram descritos na literatura, correspondendo a 1% de todos os hemangiomas. Os sintomas geralmente são inespecíficos, o que muitas vezes leva a um atraso no diagnóstico de muitos anos e que pode resultar em artropatia. O diagnóstico precoce de um hemangioma sinovial é importante, uma vez que a hemartrose recorrente pode levar a dano articular irreversível e sinovite inflamatória. Na prática, não há consenso sobre o melhor tratamento dos hemangiomas sinoviais em crianças.A ressecção total do tumor pode ser realizada por via artroscópica em formas localizadas ou de pequenas lesões, sendo necessária uma ressecção aberta associada à sinovectomia quando o hemangioma ocupa a maior parte da membrana sinovial.

Abstract

Synovial haemangioma of the knee – Diagnosis by magnetic resonance imaging

Synovial hemangioma is a rare nonneoplastic vascular malformation of the synovial membrane described by Bouchut in 1856. Fewer than 200 cases have been described in the literature, corresponding to 1% of all hemangiomas. The presenting symptoms are often non-specific, which often leads to a delay in diagnosis of many years and can result in arthropathy if left undetected. The early diagnosis of a synovial haemangioma is important as recurrent haemarthrosis may lead to irreversible joint damage and chronic inflammatory synovitis. In practice, there is no consensus on the best treatment of synovial hemangiomas in children. Total resection of the tumor can be performed by arthroscopy in localized forms and for small lesions. Open resection associated with synovectomy is necessary when the hemangioma occupies most of synovial membrane.

INTRODUÇÃO

O hemangioma sinovial é uma rara malformação vascular não neoplásica da membrana sinovial descrita por Bouchut em 1856.[1,2,3,4] Menos de 200 casos foram descritos na literatura, correspondendo a 1% de todos os hemangiomas.[1,4] Essa lesão tem predileção pela articulação do joelho e é uma causa incomum de edema crônico do joelho em crianças.[1] Também é relatado no cotovelo, punho, tornozelo, bainha do tendão sinovial e na articulação têmporo-mandibular.[2,3,4]

Os sintomas geralmente são inespecíficos, o que muitas vezes leva a um atraso no diagnóstico de muitos anos e que pode resultar em artropatia.[2] O diagnóstico precoce de um hemangioma sinovial é importante, uma vez que a hemartrose recorrente pode levar a dano articular irreversível e sinovite inflamatória.[3] O hemangioma sinovial crônico também pode resultar em erosão por pressão ou remodelação dos ossos adjacentes.[3]

Os sinais clínicos habituais são variáveis e por vezes associados: claudicação intermitente e dolorosa, limitação dos movimentos do joelho, episódios recorrentes de hemartrose e amiotrofia do músculo quadríceps.[1,2] Os hemangiomas sinoviais geralmente progridem com exacerbações e remissões, cursando com edema da articulação do joelho e dor.[4] Apresenta-se habitualmente na adolescência, com

idade média de início de 10,9 anos em meninas e 12,5 anos em meninos segundo Moon, e aproximadamente 75% dos casos se tornam sintomáticos antes dos 16 anos de idade;[2] os homens são mais afetados que as mulheres.[3]

CASO CLÍNICO

Menina de 8 anos com dor e aumento do volume do joelho direito há 5 anos, com sua mãe referindo piora gradual dos sintomas. O joelho apresenta edema de consistência amolecida à palpação, além de impossibilidade de realizar a extensão total, apresentando limitações à mobilização. Ela refere dor constante com piora após trauma, muitas vezes até ao toque e aumento do edema com mobilização (Figura 1). Refere ter realizado três embolizações prévias, duas aos 5 anos de idade e uma aos 7 anos, sem melhora da lesão.

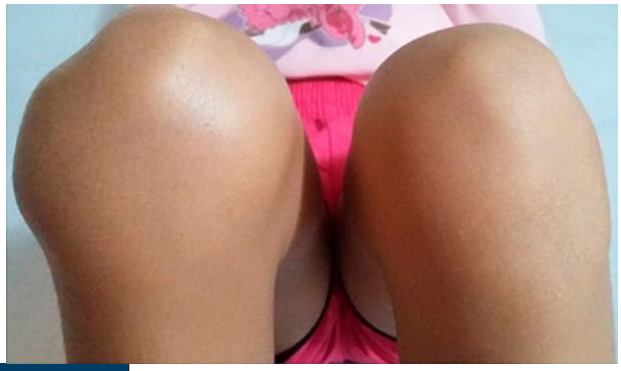

Figura 1 *Exame físico demonstra importante edema do joelho direito quando comparado com o esquerdo.*

A ressonância magnética (RM) demonstrou lesão expansiva com sinal baixo em T1 e sinal alto em T2 STIR, com áreas de baixo sinal intercaladas (flebólitos), septadas, com limites imprecisos que se expandem da gordura de Hoffa e da porção intra-articular fêmoro-tibial para a porção lateral do joelho, em contato com os côndilos femoral e tibial laterais, direcionando-se posterolateralmente na perna, em contato com a fíbula, empurrando medialmente o músculo gastrocnêmio lateral. Esta imagem mostra importante realce pelo contraste. Imagem de baixo sinal é observada em todas as seqüências estudadas adjacentes ao tecido subcutâneo lateral do joelho, sem realce pelo contraste, podendo corresponder a remanescentes de componente hemático. Os achados da imagem são compatíveis com hemangioma sinovial (Figuras 2, 3 e 4), e a paciente está em planejamento cirúrgico para ressecção da lesão desde então.

DISCUSSÃO

Os hemangiomas de tecidos moles podem ser classificados histologicamente de acordo com a natureza e o tamanho dos vasos predominantes na lesão, em subtipos

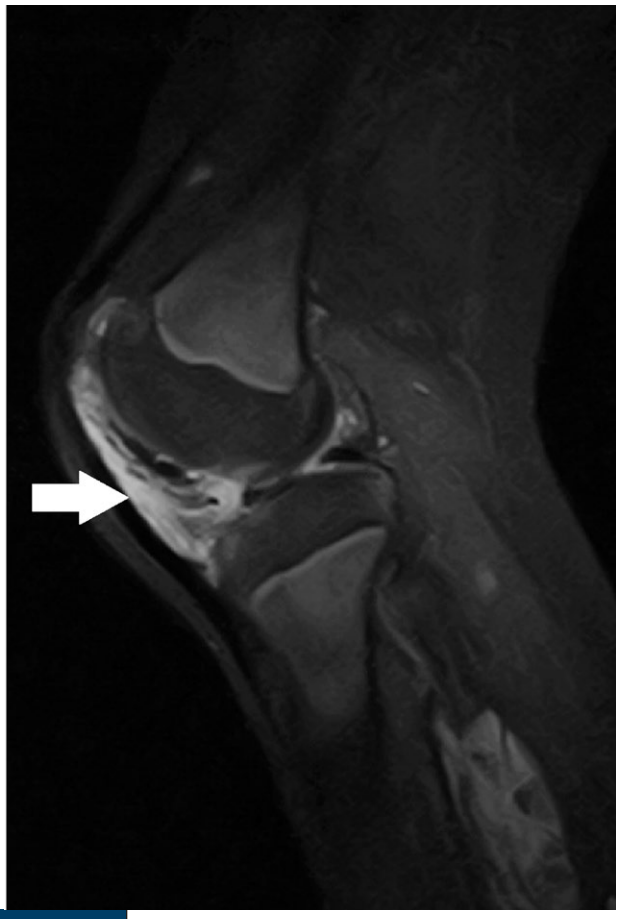

Figura 2 *RM na sequência T2 STIR no corte sagital sem contraste do joelho direito demonstrando hemangioma sinovial na gordura de Hoffa (seta branca).*

cavernoso (50%), capilar lobular (25%), arteriovenoso (20%) e venoso (5%).[2,4] Outro sistema de classificação baseia-se na relação anatômica do hemangioma com a articulação: hemangiomas justarticulares (situados externamente à cápsula articular), hemangiomas intra-articulares (situados dentro da cápsula articular) e um tipo intermediário de hemangioma (com componentes intra e extra-articulares).[2] Os hemangiomas tipos intra-articulares, bem como os intermediários, surgem da sinóvia e são denominados hemangiomas sinoviais.[2]

As radiografias são normais em mais da metade dos pacientes com hemangiomas sinoviais na articulação do joelho, mas podem demonstrar características inespecíficas, como derrame articular, massa de partes moles ao redor da articulação, osteoporose, maturação avançada da epífise e artropatia, que pode simular hemofilia.[1,2] A presença de flebólitos é muito sugestiva do diagnóstico.[1,2]

Na ultrassonografia com Doppler, o hemangioma corresponde a uma massa iso ou hiperecogênica com reforço acústico posterior, apresentando uma vascularização do tipo venosa.[1] A RM é utilizada em conjunto com a artroscopia diagnóstica em adultos, mas continua sendo o exame de escolha para a avaliação do hemangioma sinovial e estabelecer o diagnóstico e planejamento pré-operatório para a excisão cirúrgica.[1,2]

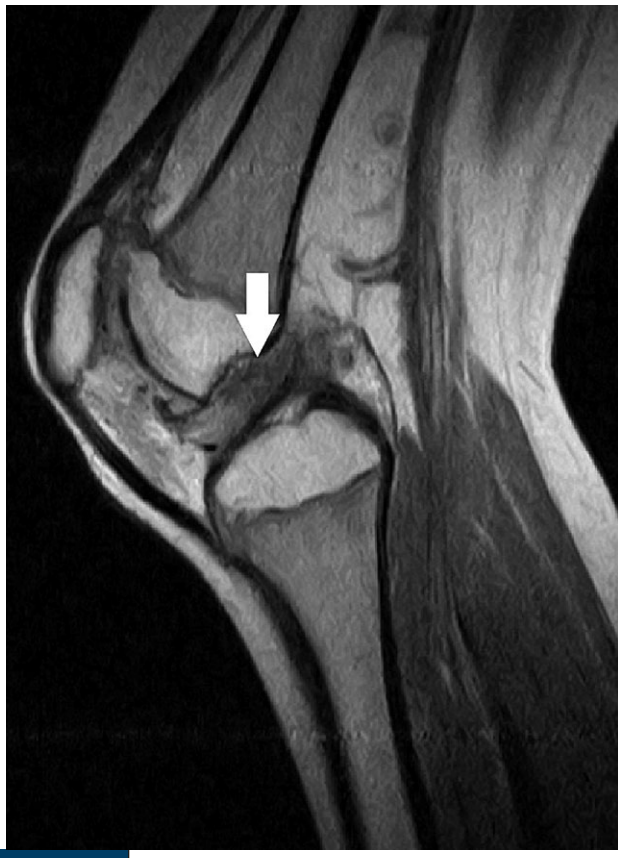

Figura 3 *RM na sequência PD no corte sagital sem contraste do joelho direito demonstrando hemangioma sinovial na gordura de Hoffa com componente intra-articular (seta branca).*

A RM geralmente mostra uma massa mal delimitada de tamanho variável, justa ou intra-articular, com iso ou hipossinal em T1 quando comparado ao músculo, um alto sinal em T2 que é realçado após injeção de gadolínio.[1] A presença de septos fibrosos ou gordurosos em seu centro são muito sugestivas, assim como as formações serpiginosas sem sinais correspondentes às estruturas vasculares.[1] Um hemangioma sinovial pode infiltrar estruturas musculares ou adiposas, ou até mesmo o osso adjacente,[1] por isso a angiografia por ressonância magnética é um método de imagem não invasivo adequado que pode ser usado avaliar a relação do hemangioma com vasos de suprimento adjacentes para melhor planejamento cirúrgico.[3]

O diagnóstico diferencial da RM traz outras patologias sinoviais, como sinovite vilonodular, osteocondromatose, hiperplasia cística, sarcoma sinovial, sinovite de artrite crônica juvenil e hemofilia.[1,2,3] O diagnóstico de certeza do hemangioma sinovial é histológico, geralmente obtido por biópsia sob artroscopia ou com cirurgia aberta.[1]

Como a RM é um exame inofensivo, ela é preferível a uma artroscopia diagnóstica, que não é desprovida do risco de lesão vascular, da cartilagem articular ou mesmo da condroepífise em crescimento em crianças com pequenas articulações.[1] Além disso, a artroscopia nem sempre permite especificar a extensão exata da lesão.[1]

O aspecto macroscópico de um hemangioma sinovial tem sido descrito como uma massa lobulada e pastosa, frequentemente corada de vermelho escuro ou marrom-mogno pela hemossiderina.[2] Ao exame microscópico, a lesão demonstra canais vasculares arborizantes no mesênquima subsinovial, que podem estar associados a

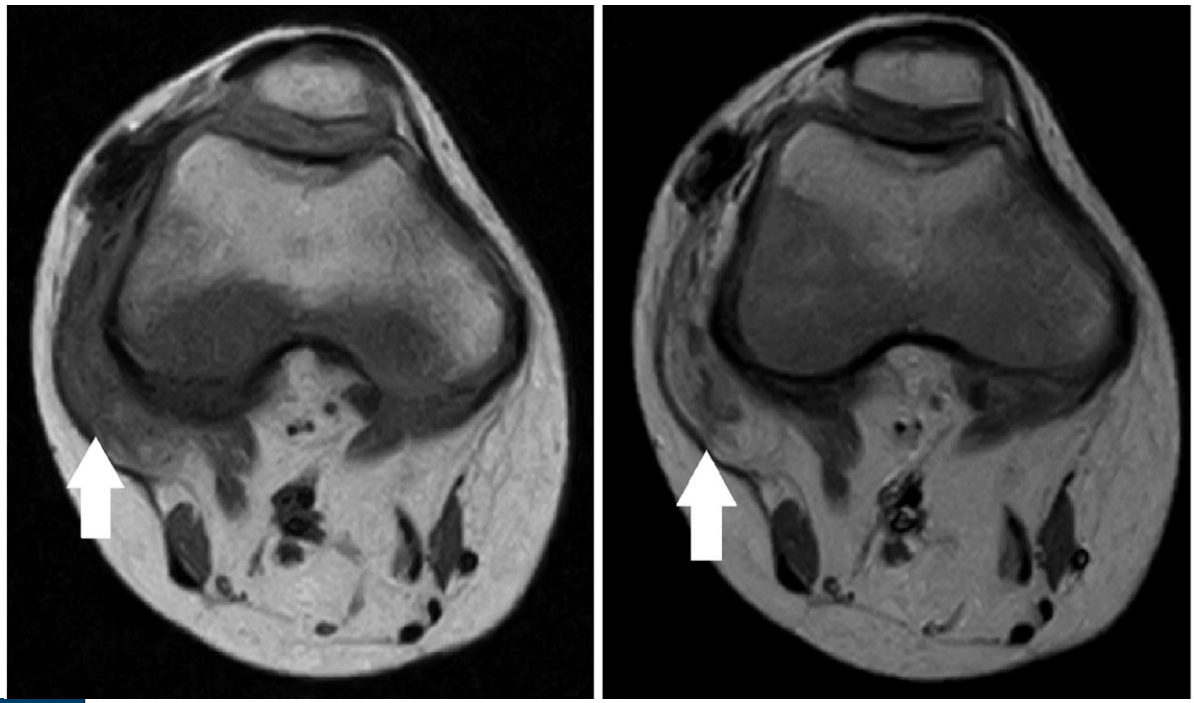

Figura 4 *Em A RM na sequência T1 no corte axial sem contraste do joelho direito demonstrando hemangioma sinovial adjacente ao côndilo femoral lateral (seta branca). Em B RM na sequência T1 no corte axial com contraste do joelho direito demonstrando hemangioma sinovial adjacente ao côndilo femoral lateral com realce importante pelo contraste (seta branca).*

deposição hiperplásica de sinóvia e hemosiderina em casos crônicos com hemartrose repetida.[2]

Em relação ao tratamento, vários métodos têm sido propostos, incluindo ressecção cirúrgica por via artroscópica ou com cirurgia aberta, tratamento por agentes esclerosantes e ablação a laser por via artroscópica.1 A embolização pré-operatória também foi utilizada para hemangiomas difusos quando a arteriografia destaca artérias tributárias de diâmetro suficiente.[1]

Na prática, não há consenso sobre o melhor tratamento dos hemangiomas sinoviais em crianças.[1] A ressecção total do tumor pode ser realizada por via artroscópica em formas localizadas ou de pequenas lesões,[1] sendo necessária uma ressecção aberta associada à sinovectomia quando o hemangioma ocupa a maior parte da membrana sinovial.[1]

REFERÊNCIAS

1. Mrani Alaoui N, Lamalmi N, Saouab R, Gourinda H, Dendane MA. Synovial hemangioma: a rare case of knee joint tumefaction in children. Ann Vasc Surg. 2014 Jan;28(1):260.e1-4.
2. Wen DW, Tan TJ, Rasheed S. Synovial haemangioma of the knee joint: an unusual cause of knee pain in a 14-month old girl. Skeletal Radiol. 2016 Jun;45(6):827-31.
3. Hoe HG, Zaki FM, Rashid AHA. Synovial Haemangioma of the Elbow: A rare paediatric case and imaging dilemma. Sultan Qaboos Univ Med J. 2018 Feb;18(1):e93-e96.
4. Arslan H, İslamoğlu N, Akdemir Z, Adanaş C. Synovial hemangioma in the knee: MRI findings. J Clin Imaging Sci. 2015 Apr 30;5:23.

LONG ABDOMINAL AORTIC STENOSIS – A CASE OF TAKAYASU ARTERITIS

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A 77-year-old female Caucasian patient with known Takayasu's arteritis diagnosed at 20 years of age was admitted to the emergency department due to diffuse sudden-onset abdominal pain. On physical examination, femoral pulses were feeble. Laboratory results were unremarkable. Abdominal CT angiography showed a long abdominal predominantly infra-renal aortic stenosis (Figures 1 and 2).

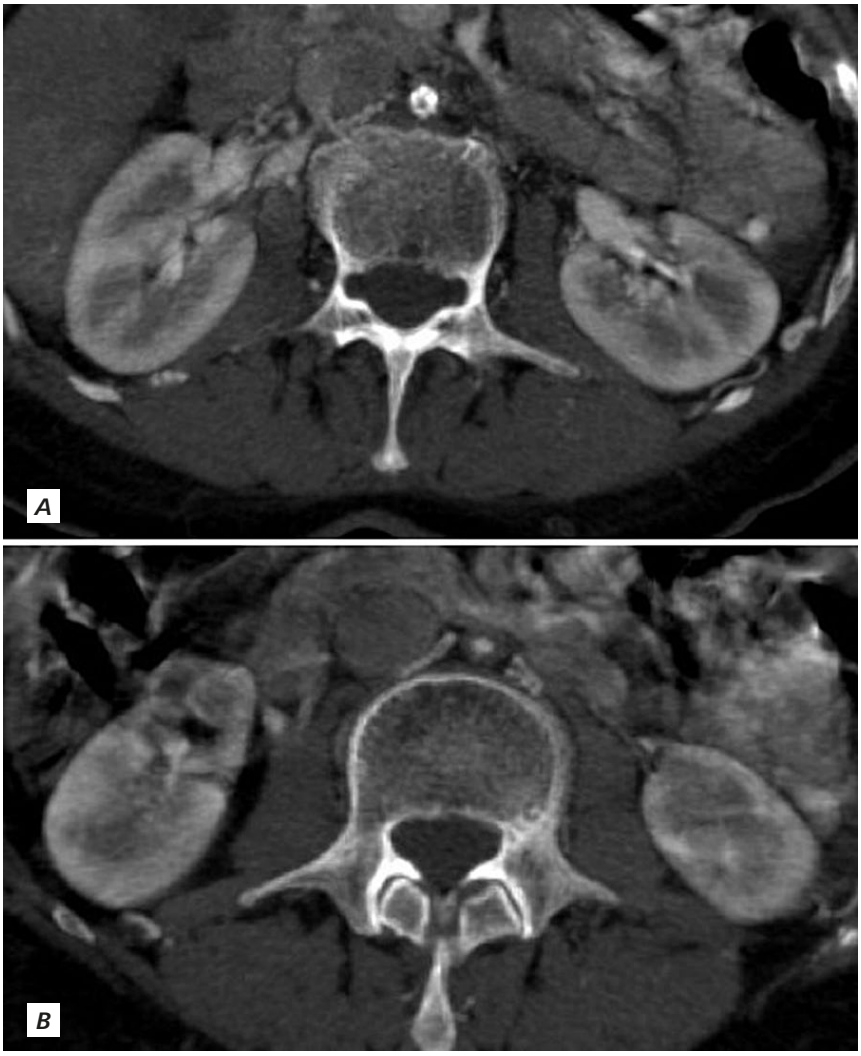

Figure 1 *MSCT angiography. Thin maximum intensity projection (MIP) axial images.* ***A*** *– markedly reduced lumen of the abdominal infra-renal aorta, with a maximum diameter of less than 1cm, with circumferential extensive calcifications.* ***B*** *– At a lower level, marked luminal reduction with peripheral and circumferential hypodense thickening of the aortic wall can be seen in keeping with the diagnosis of Takayasu Arteritis.*

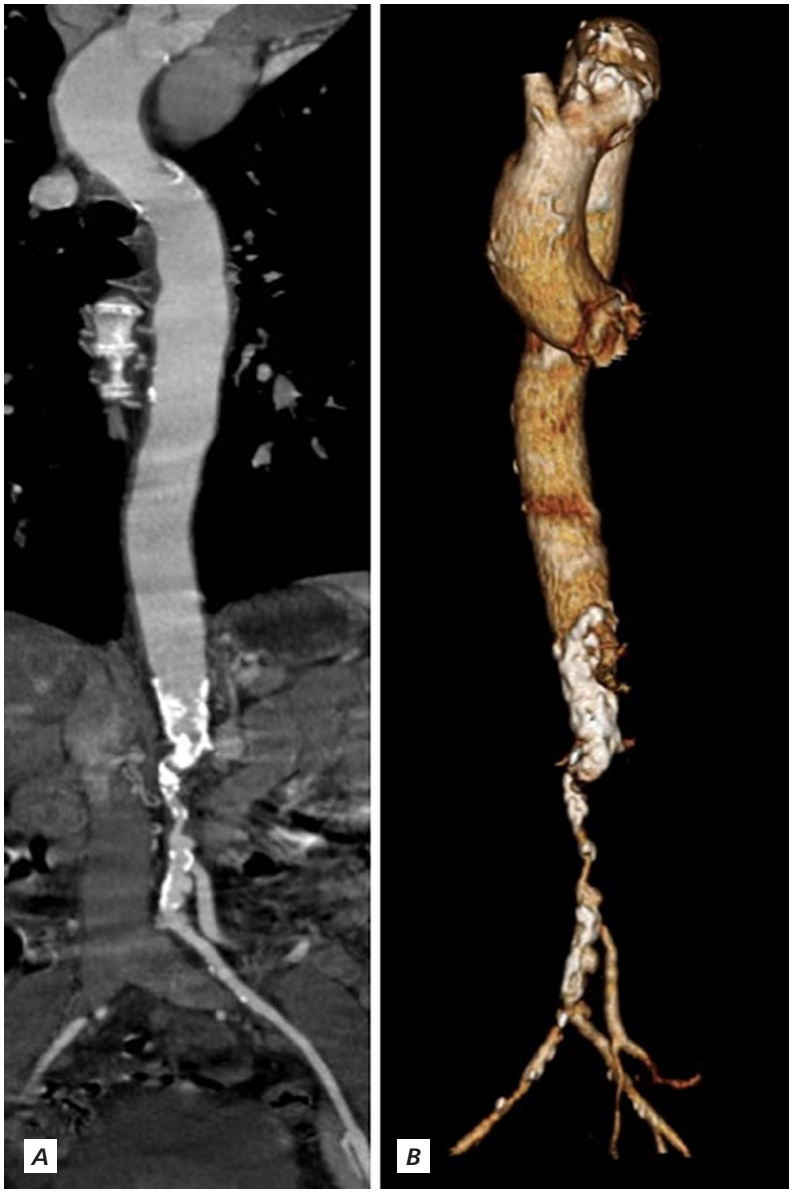

Figure 2

***A** - 2D curved reformatted thin MIP coronal image shows collateral circulation and the inferior mesenteric artery with a larger calibre than usual. **B** – 3D volume-rendered (VR) reformatted image shows the long stenosis extending from the infra-renal aorta to the iliac arteries with extensive calcifications. The large calibre of the inferior mesenteric artery and aortic arch involvement can also be appreciated.*

STERNAL CHONDROSSARCOMA

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A 28 year old woman presented a painless pre-sternal mass since childhood. At 25 years old, after pregnancy, the mass turned painful and started growing.

A percutaneous biopsy diagnosed osteochondroma (Figure 1).

She underwent partial sternectomy (Figure 2 a) and closure of the sternal defect with a Marlex mesh (Figure 2 b).

Histology revealed grade 1 chondrossarcoma with free surgical margins.

After 8 months she is free from recurrence.

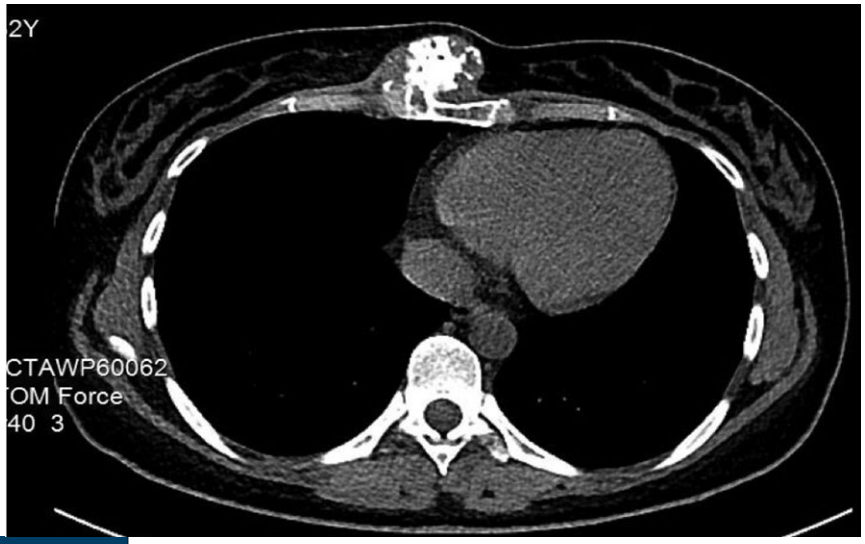

Figure 1

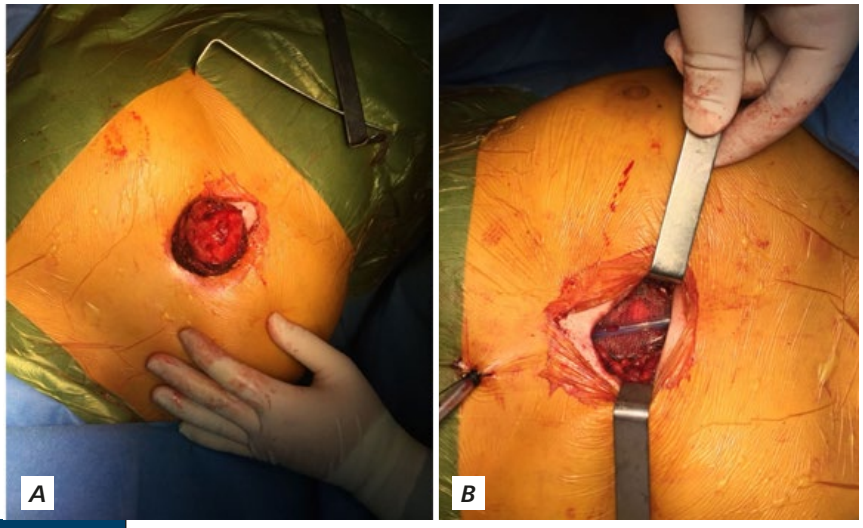

Figure 2

SURGICAL TREATMENT OF COMPLICATIONS 55 YEARS AFTER EXTRAPERIOSTEAL LUCITE BALL PLOMBAGE FOR PULMONARY TUBERCULOSIS

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In the 1930-50s, before antimicrobial drugs, collapse therapy was the mainstream of treatment for cavitary pulmonary tuberculosis.

We present an 78-year-old man with a history of pulmonary tuberculosis treated with plombage in 1962, who presented with axillary pleurocutaneous fistula (Figure 1). The patient was submitted to surgical extraction of 21 lucite balls, pleurocutaneous drainage and thoracoplasty (Figure 2).

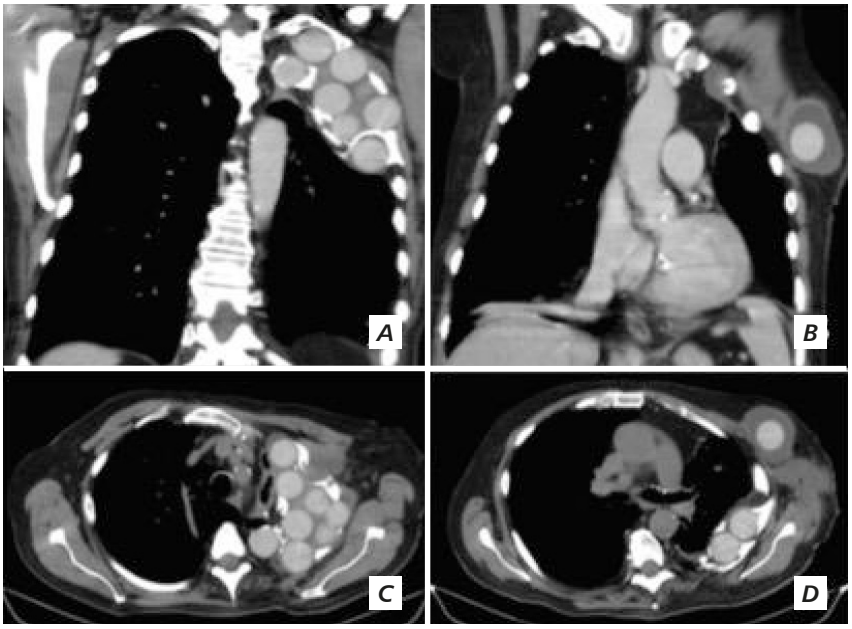

Figure 1

*CT-scan, coronal (**A** and **B**) and axial (**C** and **D**) section, with the apex of the left hemithorax filled with multiple lucite balls, each approximately 2,5cm in diameter, and extrusion of a ball into the axillary fistuluous tract.*

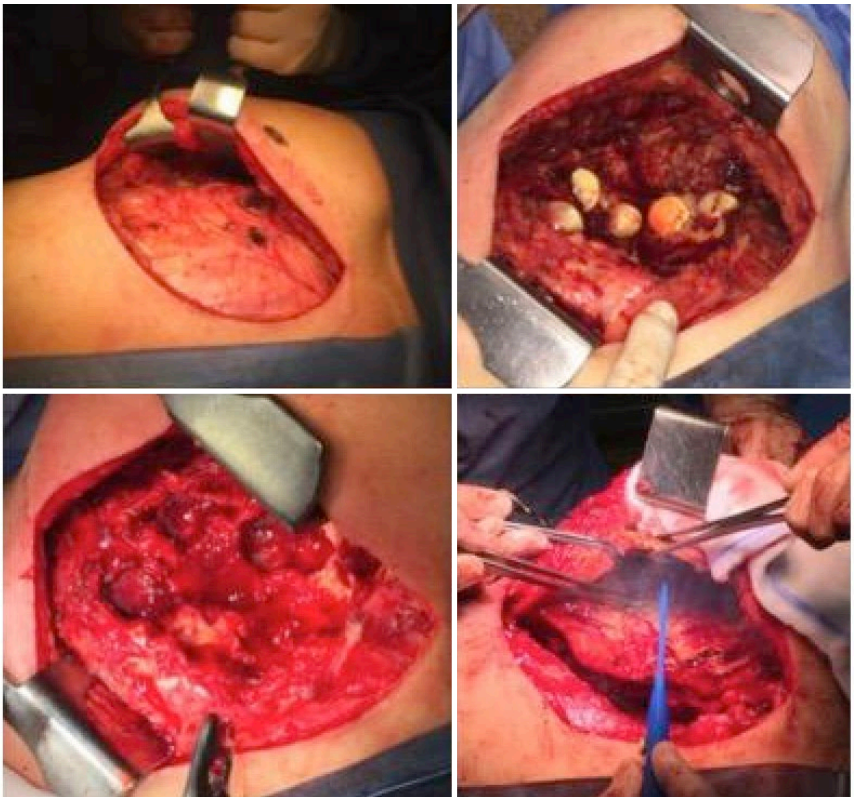

Figure 2 *Surgical extraction of 21 lucite balls, pleurocutaneous drainage and thoracoplasty (7 ribs and the tip of the scapula was remove).*

OCLUSÃO AORTO-ILÍACA E ISQUEMIA MESENTÉRICA: DIFERENTES ESPECTROS CLÍNICOS, A MESMA ENTIDADE NOSOLÓGICA

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Homem de 69 anos, hipertenso, fumador e com cardiomiopatia desenvolveu quadro de dor abdominal, retorragias e perda ponderal (10 kg), acompanhado de parestesias nos pés. Por doença arterial periférica grau III de Leriche-Fontaine e isquemia mesentérica crónica (Fig. 1) foi submetido a bypass aorto-bifemoral com extensão à artéria mesentérica superior (Fig. 2). Aos 4 anos de seguimento apresenta-se assintomático, com recuperação ponderal e bypass funcionante.

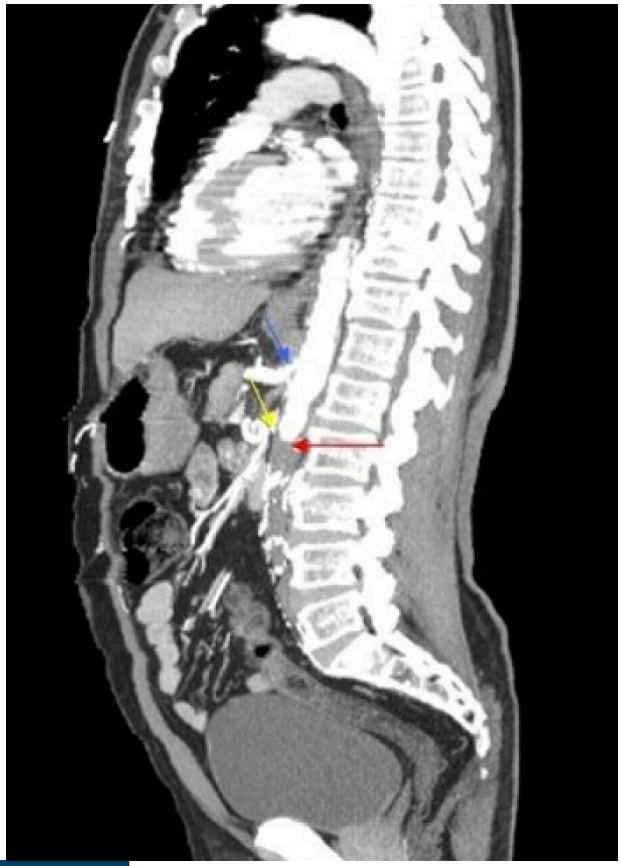

Figura 1 *TC toraco-abdomino-pélvica. Seta azul-estenose do tronco celíaco; seta amarela-oclusão curta da artéria mesentérica superior; seta vermelha-oclusão aórtica.*

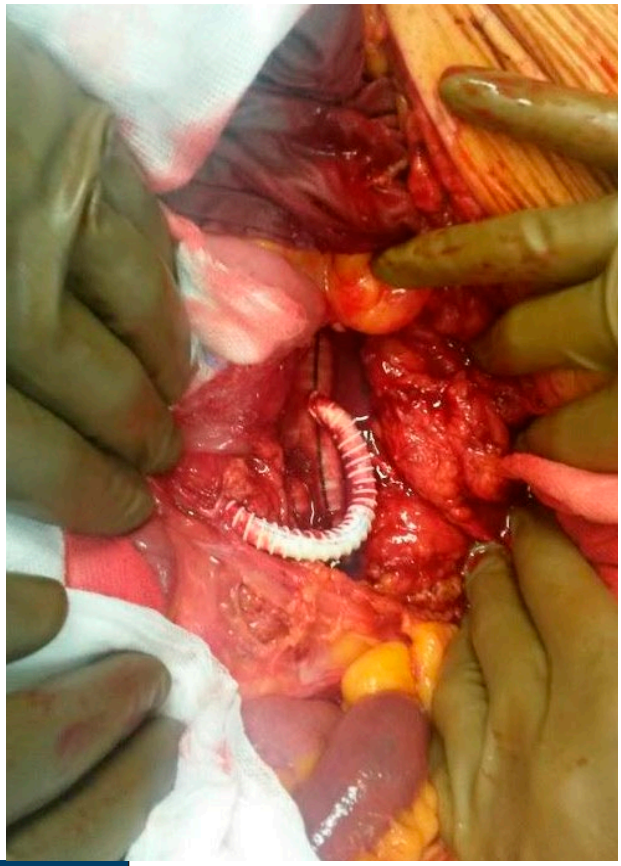

Figura 2 *Bypass aorto-bifemoral e bypass protesico-mesentérica superior.*

IATROGENIC INJURY AFTER CARDIAC SURGERY WITHIN INTENSIVE CARE CONTEXT

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65 year-old man, admitted in ICU post cardiac surgery. Pleural drains removed in postoperative. Chest radiography showed bilateral pneumothorax, apparently extensive, needing drainage, with no clinical correlation. Exam repeated in better technical conditions eliminating skin folds: it confirms small pneumothorax with no need of pleural drainage, avoiding an invasive technique.

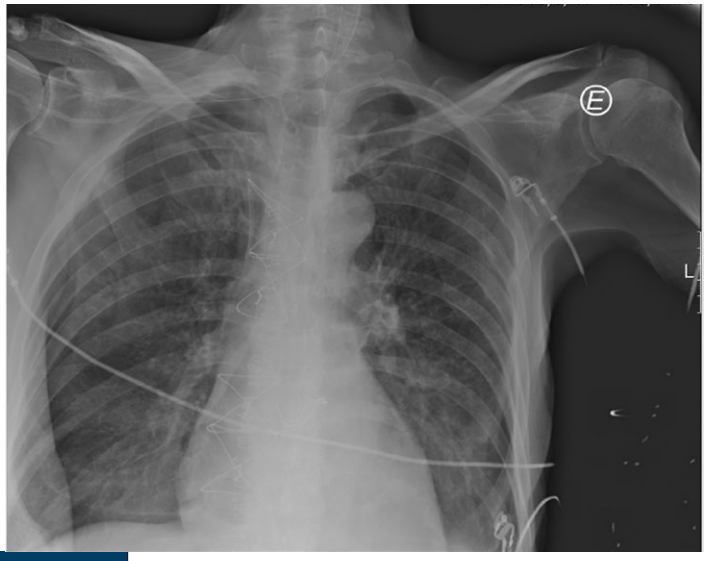

Figure 1 *Chest radiography post bilateral pleural drains removal.*

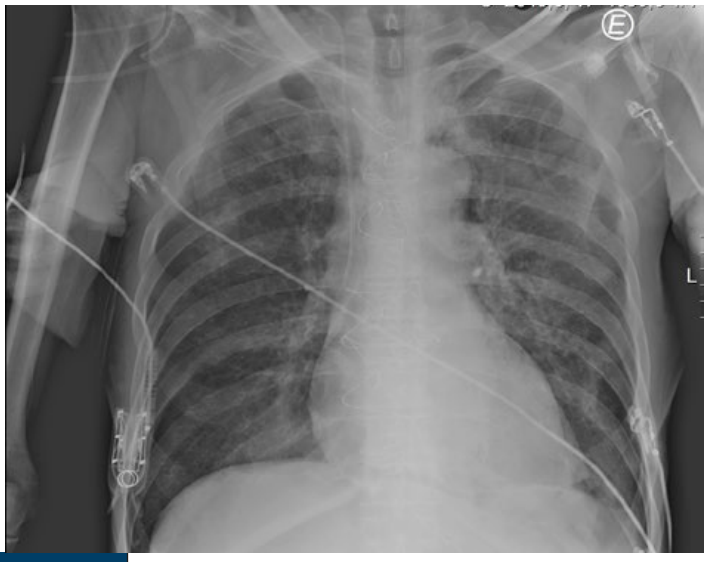

Figure 2 *Chest radiography with skin folds removal highlighting small pneumothorax.*


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