Institutional report - Congenital

Surgical treatment of anomalous coronary artery arising from the pulmonary artery

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Abstract

Anomalous coronary arteries arising from the pulmonary trunk is a rare but potentially fatal condition. We report the clinical presentation, surgical treatment and long-term follow-up of seven surgical cases of anomalous left coronary and one case of anomalous right coronary artery arising from the pulmonary artery. Age ranged from 7 months to 13 years (average: 5.09 ± 3.7 years) and weight ranged from 7 to 50 kg (average: 19.9 ± 8.8 kg). Follow-up was 100% complete: average 78 months (S.D.: 52.7 months). Direct reimplantation was the surgical technique in six cases, Takeuchi procedure in one case and subclavian artery interposition in one case. Concomitant mitral valve repair was undertaken in two cases. In two children the coronary artery anomaly was diagnosed and treated only after a first surgery for other congenital heart anomaly. Left ventricle ejection fraction was restored in those cases of pre-operative dysfunction. Mortality was not observed and all children are asymptomatic and free of reoperation.

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1. Introduction

Congenital anomalies related to cardiac coronary arteries involve a range of alterations: anomalous left and/or right coronary origin from the pulmonary artery; single ostio coronary; coronary atresia; coronary fistula; coronary aneurysm; coronary artery from the wrong aortic sinus.

After birth and the passage of desaturated blood through the pulmonary artery and the decrease in vascular pulmonary resistance, the coronaries originated anomaly from the pulmonary artery become dilated and a fistula is developed between the aortic and the pulmonary systems. Consequently an irrigation deficit for a certain region develops. This anomalous system can produce symptoms such as tiredness, angina and syncope. However, as patients generally are infants or pre-school age children, they can be misinterpreted. When not diagnosed in due time it has a poor prognosis due to the risk of acute myocardial infarction, cardiac insufficiency, arrhythmia, mitral dysfunction and sudden death. Those that develop a competent collateral system for the ischaemic region can reach adult age.

The anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare condition, occurring once in every 300,000 live newborns [1] (0.25–0.5% of heart malformations). The anomalous right coronary origin from the pulmonary artery (ARCAPA) is 10 times rarer. Altogether they represent from 6 to 27% of all congenital coronary anomalies.

ALCAPA represents the main cause of ischaemia and myocardial infarction in childhood. If not treated, it presents up to 90% mortality in the 1st year of life. They generally appear as isolated heart anomalies. Surgical treatment is always referred immediately after diagnosis, with the objective of restoring a two-coronary system.

We present the surgical experience of the Pró-Cardíaco Hospital with seven ALCAPA cases and one ARCAPA case in the period from 1995 to 2007.

2. Materials and methods

In the period from December 1995 to November 2007, seven children with ALCAPA and one with ARCAPA were submitted to surgical correction at the Pró-Cardíaco Hospital. Their ages ranged from 7 months to 13 years (average: 5.09 ± 3.7 years) and their weight ranged from 7 to 50 kg (average: 19.9 ± 8.8 kg). Two patients (25%) were <1-year-old. There was a prevalence of the male gender: 62.5% (5 patients). Follow-up was 100% complete up to 1st May 2008 with a mean time of 78 months (S.D.: 52.7 months).

The pre-surgical evaluation included heart catheterization, myocardial scintigraphy, echocardiogram and electrocardiogram. All patients repeated the echocardiogram in the immediate postoperative period and after six months of surgery.
One patient was undergoing out-patient clinic monitoring in an investigation of mitral insufficiency, with the diagnosis of anomalous coronary origin suspected due to the unusual evolution. Two other patients did undergo heart surgery on account of other anomalies without the prior diagnosis of anomalous coronary origin. One underwent a patent ductus arteriosus ligation at the age of five months with correction of the left coronary anomaly at the age of seven months. The other underwent ASD and VSD closure at the age of five months with the correction of the right coronary anomaly at the age of three years. Three cases were diagnosed during cardiac insufficiency investigation and two cases during the investigation of heart murmur.

Three patients presented an ejection fraction <30%, one patient presented an ejection fraction between 30% and 50%, and in four patients the ejection fraction was >50%.

As regards the mitral insufficiency (MI) during the pre-surgical stage, two patients presented MI +/−4+, two patients MI ++++++/+++++ and the other four did not show any MI.

The only ARCAPA case was treated with direct re-implantation. Five ALCAPA patients were treated with direct re-implantation of the left coronary ostio in the ascending aorta. One was treated through the interpositioning of a segment of the left subclavian artery between the left coronary ostio and the ascending aorta, followed by anastomosis of a PTFE graft between the aorta and the distal segment of the subclavian artery. One patient was treated with the construction of a pulmonary artery duct between the left coronary ostio and the ascending aorta (Takeuchi surgical procedure). The two patients with mitral insufficiency ++++++/+++++ were submitted to posterior annuloplasty with 5-0 Prolene suture at the same surgical moment.

3. Results

Hospital admission time ranged from 5 to 17 days (average of 8.4 ±4.1 days). The only post-surgical complication found was one case of a pericardial effusion that required surgical drainage.

There were no late deaths. All patients of the series are followed in the out-patient clinic, and post-surgical echo-cardiography study showed a preserved left ventricular function in all of them. For what concerns the mitral valve, only one patient presents mild mitral insufficiency after six months. In the long-term follow-up (12 years) the patient that underwent the Takeuchi surgical procedure presents a dilatation of the pulmonary artery duct.

In two patients the surgery for the correction of the anomalous coronary was actually a second operation (in one patient a previous sternotomy was made and in the other a previous left thoracotomy). No complications related to the redo procedure were observed.

4. Discussion

The diagnosis on the anomalous coronary origin depends of a high clinical suspicion in an infant with growth disorder, difficulty to feed, irritability, dyspnea, pale colours, and sweating associated to chest X-ray that shows cardiomegaly and an electrocardiography study that shows signs of ischaemia in the territory of the suspected coronary [2]. Older children can report tiredness in effort and chest pain. The modern diagnosis can be made with a trans-thoracic echo-cardiogram but, with the persistence of suspicion, a cardiac catheterization should be made [2].

The restoration of a two-coronary system is the objective of the surgery. Despite the common severe clinical status in preoperative presentation, surgical mortality is low (<10%) [3–5] and long-term survival is high [6]. When successful, there is a trend towards the normalization of the ventricular function in the first six months after surgery. The younger the patient at the time of surgery, the better and faster is the recovery [3]. In our series, 4/8 patients presented LV dysfunction during the pre-surgical stage. In all of them the ventricular function was re-established after six months of surgery.

The associated mitral insufficiency in ALCAPA patients has a probable ischaemic aetiology. The importance of the surgical approach to the insufficient mitral valve at the time of the coronary correction remains without a clear definition. If there is no intervention in the valve, most of the patients have the mitral insufficiency solved with the treatment of the anomalous coronary alone. A small percentage, however, will evolve with the need of replacement or valvuloplasty [5, 6]. Some uphold the approach of the mitral valve with annuloplasty whenever there is significant incompetence, and, with that, reducing its persistence in the immediate post-surgical stage [7]. We defend the mitral approach whenever there is moderate or severe insufficiency associated.

The patients in our series clearly are older than those in the majority of reports. Most patients (75%) are older than one-year-old. With that in mind we can consider them as ‘survivors’, since they probably developed a rich collateral system to the ischaemic areas and have less jeopardized myocardium. Even so, one must notice that half of them (4/8) had reduced ejection fraction that was reverted after surgery, suggesting ischaemia.

Our experience includes two cases where patients were previously submitted to surgical correction of other congenital anomalies under one year of age without the diagnosis having been done for anomalous coronary origin. In both cases there was a pulmonary hyper-flux and left–right shunt that increased the O2 concentration in the pulmonary artery and the pulmonary arterial pressure. In one of these cases the first surgery intended the surgical closure of a PDA. This patient evolved well in the immediate post-surgical period but, with the out-patient clinic follow-up, developed cardiac insufficiency symptoms. Heart catheterism showed the anomalous left coronary arising from the pulmonary artery. In a surgical approach to the PDA the access was made through a left thoracotomy, which did not allow visualizing of the coronary origin. There were no complications in the post-surgical period of this second surgery.

The second case had as first intervention the closing of an ASD and VSD. It was a right coronary originating from the pulmonary artery. We considered that the pulmonary hyper-flux associated to the left–right shunt did not allow pressure gradients between the left and right coronary...
systems for the development of fistula and consequent dilatation of the coronaries. This prevented its anatomical acknowledgement during the first surgery. After the closing of the ASD and the VSD, the patient evolved in the late out-patient clinic follow-up (after two and a half years) with signs of heart insufficiency and growth failure. A new investigation showed the coronary anomaly and the second surgery was promptly referred. Similarly to the other case, there were no complications in the post-surgical period.

Our first surgical option is always the direct re-insertion of the anomalous coronary. Only two patients were not treated with this technique. In one case the left coronary would not reach the aorta. An interpositioning of segment of the left subclavian artery was then executed with good immediate and late results. The only case of correction through the Takeuchi surgical technique is found in good long-term clinical evolution but displays moderate dilatation of the pulmonary artery duct, and requires more constant monitoring.

5. Conclusion

The surgical treatment of anomalous coronary arising from the pulmonary artery can be carried out with low morbidity/mortality even in school-age children. One should consider the diagnosis of coronary abnormalities even after successful surgical treatment of other congenital heart defects if the patient presents an unfavourable evolution.

References