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Total Anomalus Pulmonary Venous Connection: About a Case Operated on at the Andre Festoc Center of Bamako

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Abstract: Total anomalus pulmonary venous drainage refers to the absence of connection of the four pulmonary veins to the left atrium. This anomaly is often associated with atrial septal defect This is a rare malformation, with total forms accounting for 0.4% of all congenital heart defects, while partial forms account for between 0.6 and 0.7%. Diagnosis is a delicate matter, made by cardiac ultrasound performed by an experienced operator. The aim of this work was to establish the characteristics of this pathology, to determine the operative indications, and to evaluate the operative results. We report the case of the first complete cure of total anomalous pulmonary venous return at the Festoc center in Bamako. The patient was 08 years old and was initially referred to us for management of a sinus venosus atrial septal defect with symptoms of precordialgia and stage 2 to 3 dyspnoea. Examination revealed a mesocardial systolic murmur. Cardiac ultrasound revealed a 38 mm wide atrial septal defect sinus veinosus with no superior vena cava. Surgical exploration revealed a single atrium with total abnormal pulmonary venous return. Given the local team's lack of experience with this complex congenital anomaly, a collegial decision was taken to enlarge the ASV. Three weeks later, the patient underwent a repeat operation under extracorporeal circulation after an angioscan scan had revealed the collector. Surgical exploration revealed a single atrium with a small left atrium. The collector ended at the TVI. We re-implanted the collector in the left atrium and ligated the TVI. Pulmonary venous return anomalies are rare conditions and represent a heterogeneous group of cardiovascular malformations. Their incidence is estimated at 0.4-0.7% of all congenital heart defects. Their management requires a certain level of logistical organization and technical expertise, which can be envisaged in developing countries. North-South cooperation is also a very important aspect in the ongoing training of medical staff and the management of certain complex cardiopathies.

Keywords: Anomalous, Pulmonary Venous, Repair, Bamako

1. Introduction

Total anomalous pulmonary venous connection refers to all congenital malformations of the heart involving anomalies in the connection of the pulmonary veins to the left atrium. Depending on the number of veins involved, total anomalous pulmonary venous return is defined as none of the four pulmonary veins terminating in the left atrium. This anomaly is often associated with atrial septal defect. [1] This is a rare malformation, with total forms accounting for 0.4% of all congenital heart defects, while partial forms account for between 0.6 and 0.7%. Diagnosis is a delicate matter, made by cardiac ultrasound performed by an experienced operator. Curative treatment is surgical, with the aim of providing an unobstructed route for pulmonary venous drainage into the left atrium, closing any residual atrial septal defect and eliminating all other pulmonary venous drainage routes. [1]

2. Observation

This is an 08 year old girl, 1st of 03 siblings, from a full term pregnancy and eutocique vaginal delivery who was initially referred to us for management of a sinus veinosus AIC with symptoms that had been evolving for 1 year consisting of precordialgia and dyspnoea stage 2 to 3.

On physical examination, the patient was in good general condition with a normal facies and a pulse ox of 98%. There was no cyanosis or digital hippocratism.

The chest X-ray showed cardiomegaly associated with pulmonary hypervascularisation.

The electrocardiogram showed right atrial hypertrophy.

Cardiac ultrasound revealed a 38 mm wide sinus venosus atrial septal defect with no superior vena cava. In addition, there was a 4 mm pericardial detachment latero VG and 5 mmm opposite the right ventricle. The size of the annulus and the pulmonary branches were normal according to this first cardiac ultrasound. There were no other abnormalities. Apart from an anaemia of 9 g/dl, the entire preoperative work-up was normal.

She therefore underwent an initial operation during which a vertical median sternotomy was performed, followed by placement of extracorporeal circulation between an aortic cannula and 2 vena cava cannulae. After clamping and right atriotomy, surgical exploration revealed a single atrium with total abnormal pulmonary venous return. Given the local team's lack of experience with this complex congenital anomaly, a collegial decision was taken by the surgeons, anaesthetists and cardiologist to widen the atrial septal defect and defer a complete cure.

A thoracic angioscan was performed, which showed significant dilatation of the innominate venous trunk and superior vena cava. The vertical vein was also identified (Figures 1 and 2).

Three weeks later, the patient underwent a repeat operation with a team of cooperating surgeons and natives, under extracorporeal circulation after an angioscan had been performed, which revealed the collector where the 4 pulmonary veins terminated (Figures 1-2). After stereotomy and release of adhesions.

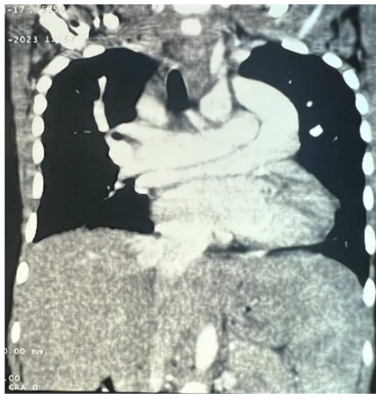

Figure 1. *Thoracic angioscan frontal section showing significant dilatation of the TVI and vertical vein.*

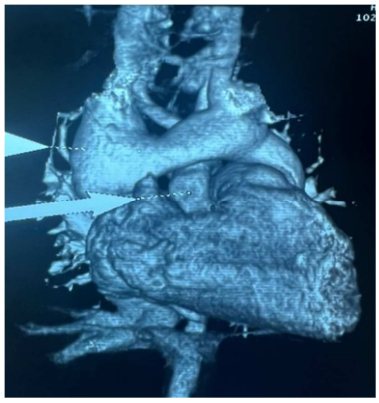

Figure 2. *Thoracic angioscanner frontal section showing significant dilatation of the TVI and vertical vein and the collector.*

Surgical exploration revealed a single atrium with a small left atrium (Figure 3). The collector ended at the level of the vertical vein (Figure 7).

We performed a complete dissection of the collector and left atrium, then clamped and longitudinally opened the collector for approximately 7 cm, before anastomosing it with the posterior surface of the left atrium (Figure 4). The interatrial septum was restored with a large patch (Figures 5-6).

The vertical vein was then ligated (Figure 7). The clamping time was 73 minutes, with a bypass time of 73 minutes and an assistance time of 31 minutes.

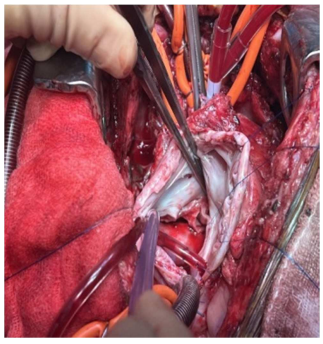

Figure 3. *(Intraoperative view): Opening of the lateral wall of the left atrium through the interatrial septum.*

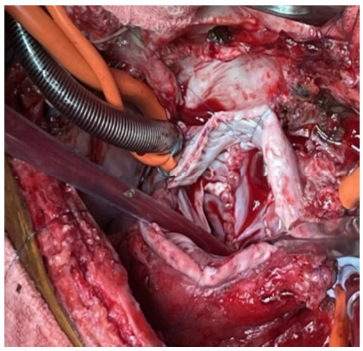

Figure 4. *(Intraoperative view): Suturing the collector to the posterior surface of the left atrium.*

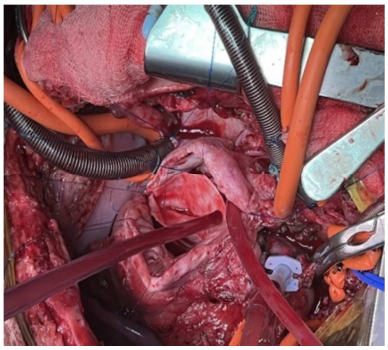

Figure 5. *(Intraoperative view): inter-auricular patch suture.*

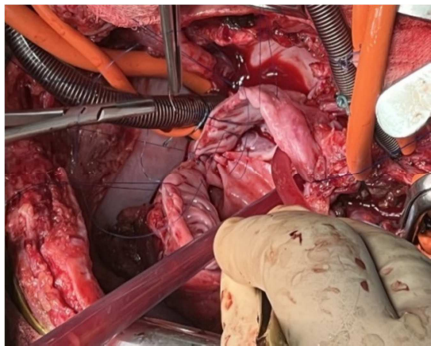

Figure 6. *(Intraoperative view): Opening of the lateral wall of the left atrium through the interventricular septum. Auriculaire.*

Post-operative management was straightforward. The drains were removed at day 3 post-op. She was discharged from intensive care four days after the operation and from hospital seven days later.

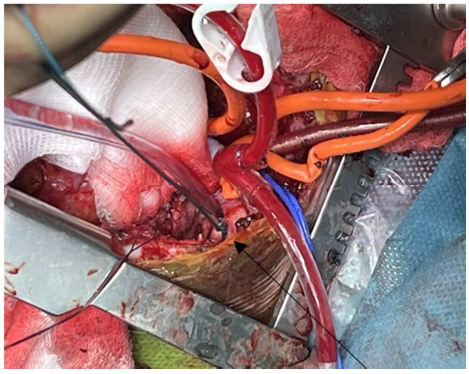

Figure 7. *(Intraoperative view): Vertical vein ligation.*

3. Discussion

Total anomalous pulmonary venous connection account for 1-3% of congenital heart disease. Isolated forms account for two-thirds of cases. The other third usually accompanies complex heart disease with right isomerism [1].

The incidence of total anomalous pulmonary venous connection is estimated at 0.4-0.7% in autopsy series [2]. The prevalence was estimated at 0.2% in a recent retrospective study [3].

This demonstrates that total anomalous pulmonary venous connection is a rare congenital heart disease.

The distribution is the same in both sexes, except for subdiaphragmatic pulmonary venous returns, which are more frequent in males. Our patient was female.

In fact, several authors note a male predominance. [5-7]. A few rare familial cases have been reported, but the malformation is usually sporadic.

According to a study by E. Robert-Gnansia et al [9], inversion-duplication of chromosome 22q-11.2 is associated with 40% of pulmonary venous return anomalies.

Clinical signs depend on the permeability of the atrial septal defect (ASD) and on the presence or absence of obstruction to the return of pulmonary venous blood to the heart [11]. In our patient, dyspnoea was predominantly NYHA stage 3.

Poorly tolerated, early-onset forms are often associated with the presence of venous return obstruction [12].

Ultrasound is the imaging modality of choice for the positive diagnosis of total pulmonary venous return. On cardiac ultrasound, the right cavities are dilated, the left atrium is small, and the pulmonary veins do not drain into the left atrium. These signs prompt a search for a collector, which may be supra-, infra- or intracardiac.

Unblocked RVPAT in older children is corrected by bypass surgery and conventional myocardial protection. They pose no particular problems. Blocked total anomalous pulmonary venous connection needs to be corrected with bypass surgery and circulatory arrest in deep hypothermia.

The surgical mortality of RVPAT is reported to be less than 5-10%, but reaches 20% in patients with severe pulmonary hypertension [13].

Post-operative mortality in large published series ranges from 5 to 20%, but recent series report very low mortality (2%) in specialized referral centers [13].

4. Conclusion

Total anomalous pulmonary venous connection are rare pathologies and represent a heterogeneous group of cardiovascular malformations. Their incidence is estimated at 0.4-0.7% of all congenital heart defects. Their management requires a certain level of logistical organization and technical expertise, which can be envisaged in developing countries. North-South cooperation is also a very important aspect in the ongoing training of medical staff and the management of certain complex cardiopathies.

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Biography

Baba Ibrahima Diarra, thoracic and cardiovascular surgeon, hospital practitioner, head of the cardiovascular surgery department at the "Luxembourg" mother and child hospital in Bamako.