

The Management of Left Pulmonary Artery Sling in Senegal: Our Experience from One Case

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Abstract:

Key words: Pulmonary Artery Sling (PAS) is a rare congenital heart disease due to abnormal origin of Left Pulmonary artery, Pulmonary Artery (LPA) from the Right Pulmonary Artery (RPA), then encircling the distal part of the trachea. The treatment is a surgical repair consisting of reimplantation of the LPA on the main pulmonary artery over cardiopulmonary bypass. The authors report a clinical case of PAS in a 5.5 years old boy diagnosed in Senegal then successfully operated in France.

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Introduction:

Pulmonary artery sling is a rare congenital heart disease in which the LPA is originating from the RPA then encircling the distal trachea and the right main bronchus.¹ This is a malformation that is often revealed while investigating respiratory distress and congenital heart disease in infants. The respiratory signs are not specific and may include wheezing, stridor, dyspnea on effort, coughing and recurrent infections of lower airways.² In ideal situation,³ the diagnosis is made by echocardiogram while the bronchoscopy accesses the bronchial stenosis. The treatment is strictly surgical and classically consists of cutting the LPA then reimplanting it on the pulmonary artery (PA) main stem by sternotomy and cardiopulmonary bypass (CPB). Tracheal repair is indicated if the patient is clinically symptomatic. We report a case of PAS in a 5.5 years old boy diagnosed in the Department of Cardiology of the University Hospital of Fann Dakar then successfully repaired in Monaco.

Case report:

AD, a boy of 5.5 years old was admitted for dyspnea with stridor occurring on exercise. There was no dizziness, sweating or cyanosis. On the previous medical history, a recurring bronchitis is reported but there was no consanguinity and the pregnancy

was followed up to deliverance. He was hospitalized twice at the ages of 2 months old and 3.5 months old respectively for dyspnea and stridor that were treated by salbutamol puff and physiotherapy. The physical examination showed a good growth regarding the weight (21 Kg) and height (121cm) which were corresponding to Z score -0.94. The cardiovascular examination was normal with oxygen saturation at 98%. The respiratory examination showed suprasternal retraction and expiratory wheezing. There were no abnormal findings in the liver, ENT and dental areas. On the chest X rays, cardiac outline, pulmonary



Fig.-1: Lateral view of chest X ray.

parenchyma, vessels, and pleura were normal. On the lateral view (figure 1) it was difficult to spot a tracheal stenosis as it was not quite penetrated.

The echocardiogram found situs solitus levocardia and good size of cardiac chambers. The cardiac function was good and the aortic arch was also normal, left-sided and free of abnormality. There was no coarctation or interrupted aorta. The PA stem had a good size with a normal RPA but the LPA was not well detected. On the angio CT, the LPA had a retro tracheal tract with a severe stenosis on the tracheal segment followed by a normal size (Fig 2). The PA stem and the RPA were normal and there was no significant tracheal stenosis. The lab tests were normal.

We indicated a surgical repair and AD was transferred to the cardiothoracic centre of Monaco where he underwent reimplantation of the the LPA over CPB and beating heart. LPA was cut at its origin, running suture of the stump and switch of the LPA from the bronchus. Then the LPA was shortened and reimplanted on the main PA. The CPB duration was 29 minutes. The postoperative echocardiogram (Fig3) showed good function of both right and left ventricles and no obstacles on their outputs tracts. The RPA was seen in a position just little far than usual but there was no tricuspid regurgitation and the atrial septal was normally curved. The postoperative course was simple and the patient discharged from hospital at day 7.

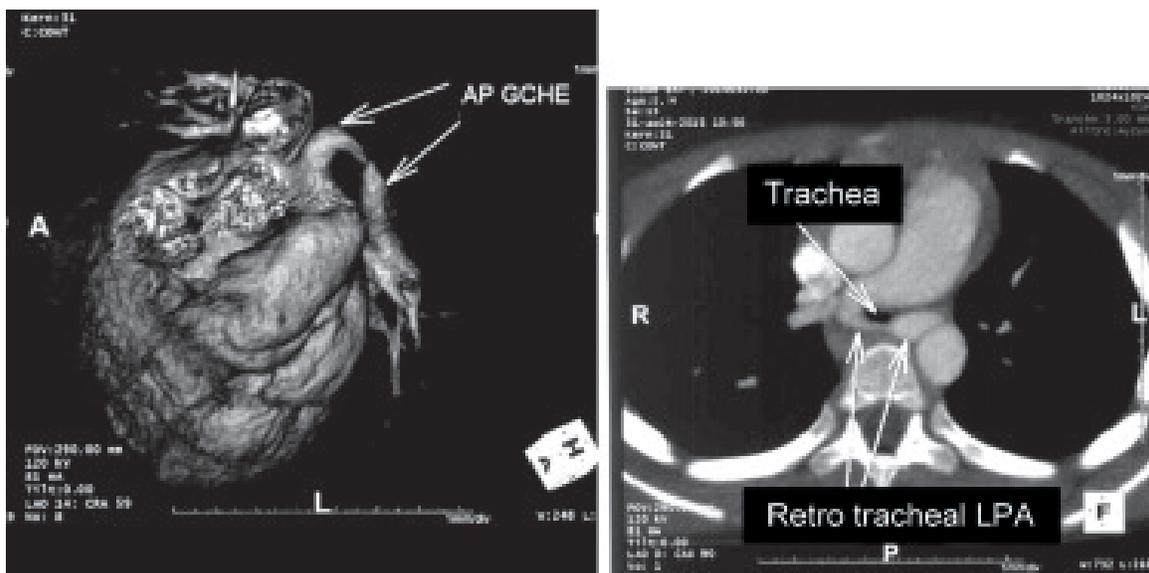


Fig.-2: Chest CT scan: retro tracheal LPA visible tracheal stenosis

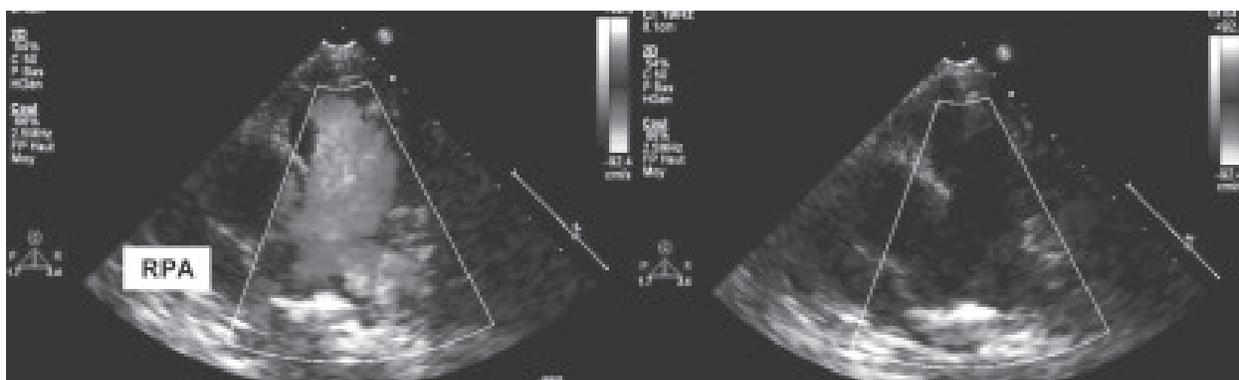


Fig.-3: Post operative echocardiography (flow going from the MPA to the LPA and the origin of RPA. No aliasing.)

Discussion:

Retro tracheal LPA is a rare congenital malformation that happens at the 5th week of pregnancy and that represents about 3% of the etiologies of extrinsic tracheal stenosis according to Landing.⁴ In the literature, the PAS is often described as asymptomatic during childhood. Nevertheless when the parents of patients are further interviewed, they report some signs. These may include dyspnea, coughing, wheezing, stridor and limitation of exercise.^{2,3,5,6} Likewise our patient, the male sex was predominant in many studies such as the series of Yu and Gikonyo who reported 72% and 60% of male patients respectively.^{2,5} Although there are 3 theories to explain this vascular malformation, none is unanimous. The first one state it is a total absence of development of the LPA and the blood supply comes from the RPA. For the 2nd theory it is rather an anomalous implantation of the LPA and the primitive vascular pedicle of lung. The last and most recent theory claims it is a persisting connection between the primitive pulmonary vascular pedicles. All of these theories are accepted and each is validated depending on the type of implantation of the LPA.⁷ Diagnosis is usually confirmed by CT scan and RMN that show both anomalous PA and tracheal compression; bronchoscopy is useful to assess respiratory tract.⁸

Once the diagnosis is made, the treatment is a surgical repair. For our patient a reimplantation of the LPA by median sternotomy over CPB without cardioplegia was performed. No tracheal repair was performed. Indeed median sternotomy is the classical approach to getting to the heart in many series. Left thoracotomy is another alternative. For instance regarding the surgical approach, Barker et al.⁹ have reported 67% of median sternotomy and 33% of thoracotomy in a series of 24 children operated for PAS. The main reason is that sternotomy makes easier the CPB and allow a good access to the PA and its branches in their intra pericardial part. Tracheal repair is only indicated in case of significant tracheal stenosis.¹⁰ The operative time varies depending on association with other malformations or not. The immediate post-operative course was good allowing hospital discharge after a week. Most of authors emphasize the low rate of morbidity and mortality for PAS.^{4,6,9} To illustrate, in the series of Young, no death was noticed in 9 cases of LPA reimplantation for PAS.¹⁰

Conclusion

Because of the no-specific signs of pulmonary artery sling and its rarity, this malformation is often not diagnosed at the clinical assessment. Since cardiopulmonary bypass is used, morbidity related to reimplantation of left pulmonary artery to correct the condition has been significantly decreased.

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