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Complete repair in a two day baby with Valvular Pulmonary Atresia.

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Abstract- *Two days old baby was diagnosed to have Valvular Atresia, intact septum, severe TR, Severe PAH. PDA was supplying the branch Pas. This baby was born in Dr YS Parmar Government Medical college Nahan, HP. Baby was transferred to Artemis hospital Gurugram. It was 4 hours journey.Transannular RVOT Patch (Autologus Pericardium), PDA Ligation, RV Overhaul, TV Repair, PFO Narrowed was done . Baby was discharged home after 7 days. And thriving*

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I. Hospital course

Presenting Complaint: Cyanosis,Respiratory distress

History of Presenting Complaint: Baby 2 days old male child, 2nd birth order from Himachal, is a product of full term normal vaginal delivery. Baby was suspected to have heart disease soon after birth during evaluation of cyanosis and referred to Artemis as a case of duct dependent congenital heart disease on day 2 of life. There was no h/o seizures, unconsciousness or family history of congenital heart disease. Examination: Alert, afebrile, Vitals within normal limits.Saturation in room air 65%. All peripheral pulses well felt. Cyanosis ++, no pallor/ icterus/pedal edema or facial puffiness.CVS: No Cardiomegaly,S1- Normal, S2 single.Systolic murmur Gr 2/6 at left infraclavicular region. RS: Air Entry Bilaterally Equal; No added sounds,Rest of the systemic examination was normal Hospital Course: At admission in ICU baby had respiratory distress, SpO2 74%. Prostaglandin infusion started at admission and air by nasal prongs started. Sepsis screen was negative at admission. Baby improved on prostaglandin infusion and after stabilisation was taken for surgery on day 5 of life.Admission ECHO: Pulmonary atresia (Membranous), Severe TR, Small ASD, Large PDA, Good size confluent Branch PAs, Bovine ARCH and normal LV systolic function.

He underwent Transannular RVOT Patch (Autologus Pericardium), PDA Ligation, RV Overhaul, TV Repair, PFO Narrowed. The risk and benefit of the surgery was explained in detail to the parents and the possibility of prolonged ventilation and ICU stay was also explained in detail.

Details of Procedure

Bypass Time : 139 mins Cross-clamp time : 67 mins Technique : Drains: Two (12 Fr straight, 16 Fr straight)
Peritoneal Drain: 12 FrPacing Wires: Two Atrial, Two Ventricular , Incision: Median Sternotomy, Cannulation: Bicaval (SVC-12 Fr angled, IVC- 12 Fr Angled), Ascending Aortic (8 Fr DLP)
CPB: Complete (SVC, IVC snared) , Myocardial Protection: Cold Antegrade Blood Cardioplegia (Delnido), Topical cooling with ice water

II. Procedure:

Following a median sternotomy, Thymus was removed to improve exposure. Pericardium was opened and hitched to the sternum using 2-0 silk stays. Hugely dilate RA, Small RV with Severe RVH, adequate sized MPA were noted. CPB was established with SVC, IVC and ascending aortic cannulation within 6-0 prolene purse strings, all cannulas were de-aired and connected to the respective limbs of the CPB circuit. A cardioplegia needle was inserted in the ascending aorta, also de-aired and connected to the plegia circuit. PDA was closed with a medium ligaclip. CPB was established with good return and flows. Systemic cooling was up to 26°C. LV was vented through a vent inserted in the left superior pulmonary vein within a 6-0 prolene purse string. Ascending aorta, MPA, RPA and LPA were mobilised on CPB.With CPB flows transiently reduced, aorta was cross clamped and myocardial arrest was achieved with cold, blood based cardioplegia delivered ante grade through the aortic root. SVC, IVC were snared and a right atriotomy was performed. Tricuspid valve is thickened with thick short chordae. Tethered chords of Anterior and Septal leaflets were sharply split to increase length.RV muscle in the apical RV were divided sharply to increase RV capacity.A large PFO was present and has been narrowed to about 4 mm using 6-0 prolene sutures

A vertical pulmonary arteriotomy was performed on the MPA, between stay sutures and extended proximally up to the Pulmonary annulus, thick plate of tissue at the annulus has been excised and the incision was extended over the RV infundibulum, Thickened RV muscle in the infundibulum was cored out. RVOT and MPA were reconstructed with a Autologous (Transannular) Pericardial patch and using 7-0 Prolene continuous sutures.
Patient was rewarmed to 36 °C. LA was de-aired and the aortic root was vented. With the head end in steep Trendlenburgh position, aortic root and LV vent on suction, flows were transiently reduced and the aortic cross clamp was removed. Heart spontaneously returned to sinus rhythm. With the patient rewarmed to normothermiaand on inotropic support, he was easily weaned off bypass and LA vent was removed. About 250 cc of ultra-filtrate was removed post bypass, using 'Modified ultrafiltration'. Following reversal of heparin by protamine, all cannulas were removed; purse strings were tied down and reinforced with 6-0 sutures.Following hemostasis, sternum was left open, skin cover has was provided with a silicon patch sutured with 5-0 prolene (over 2 chest drains- right pleural and mediastinal and over 4 pacing wires 2 atrial and 2 ventricular). 12 Fr Peritoneal drain was inserted for abdominal decompression

Post operative course: He was transferred from OT with open chest and on high inotropic support (adrenaline, dopamine, levosimendan). Chest was closed on POD 1. Failed Extubation attempt on POD3 because of severe laryngeal edema. He was extubated on POD5 to oxygen by nasal cannula. Ionotropes were tapered off by POD7.
At discharge he was hemodynamically stable, saturations are 90% on room air and is accepting full feeds Significant Medication Given: Adrenaline, dopamine, levosimendan, meropenem, colistin, lasix, aldactone, aminoven. Condition at Discharge: He was hemodynamically stable, afebrile and his actiauscultation were good. Nowhe is feeding well. Saturation in room air is 90 % and his chest is clear on auscultation. Wound is healthy and healing well and sternum is stable.

Follow up Echo :- wide open RVOT, confluent PA, free PR, moderate TR (PG 27 mmHg), normal LV systolic function, RV diastolic dysfunction, PFO bidirectional, no pericardial, pleural effusion

III. Discussion

Pulmonary atresia is a form of heart disease in which the pulmonary valve does not form properly. It is present from birth (congenital heart disease). The pulmonary valve is an opening on the right side of the heart that regulates blood flow from the right ventricle (right side pumping chamber) to the lungs.

In pulmonary atresia, the valve leaflets are fused. This causes a solid sheet of tissue to form where the valve opening should be. Normal blood flow to the lung is blocked as a result. Because of this defect, blood from the right side of the heart is restricted from reaching the lungs to pick up oxygen.

Causes

As with most congenital heart diseases, there is no known cause of pulmonary atresia. The condition is linked with another type of congenital heart defect called a patent ductus arteriosus (PDA).
Pulmonary atresia may occur with or without a ventricular septal defect (VSD).

1. If the person does not have a VSD, the condition is called pulmonary atresia with intact 7ventricular septum (PA/IVS).
2. If the person has both problems, the condition is called pulmonary atresia with VSD. This is an extreme form of tetralogy of Fallot.

Although both conditions are called pulmonary atresia, they are actually different defects. This article discusses pulmonary atresia without a VSD.
People with PA/IVS may also have a poorly developed tricuspid valve. They may also have an underdeveloped or very thick right ventricle, and abnormal blood vessels feeding the heart. Less commonly, structures in the left ventricle, aortic valve, and right atrium are involved

Types-

Atresia of the pulmonary valve classifies into membranous and muscular forms. It is essential to distinguish between these two types, as a membranous form of PA has better long term prognosis when compared to muscular PA due to the higher incidence of abnormal connections (discussed in the latter part of this section) between the RV and coronary arteries. Due to high pressures in the RV, the tricuspid valve is generally abnormal. The tricuspid valve can be hypoplastic, dysplastic, and can have malformed chordal apparatus.

Another characteristic feature, if present, of PA-IVS, are the abnormal connections between the RV and coronary arteries. The RV, especially in patients with a competent tricuspid valve, is hypertensive due to lack of egress for the blood. Due to which the RV develop these abnormal connections with the epicardial coronary arteries, which help to decompress the ventricle. These abnormal connections, when present over time, leads to progressive stenosis of the coronary arteries related to high-velocity blood flowing through these abnormal connections. Due to progressive stenosis of the coronary arteries over time, some parts of the myocardium

depends on right ventricle for the perfusion and is known as right ventricular dependent coronary circulation (RVDCC). Due to the progressive nature of RVDCC, it correlates with poor prognosis.

Hemodynamics

The hemodynamics can differ greatly depending on the specific constellation of anatomic variance. There will be progressive cyanosis in the early neonatal period. They will typically be a murmur related to the degree of tricuspid valve regurgitation as well as a ductal murmur. These patients will need prostaglandin infusion in the neonatal period to ensure patency of the patent ductus arteriosus in order to maintain adequate pulmonary blood flow

IV. Conclusion

Pulmonary Atresia is a nightmare. Surgery is very difficult. Post operation course in storming. Transportation to a specialized pediatriccardiac surgery unit is also challenging.

But the above case is a great teaching for the future.

Duct dependent lesions can be transferred without prostaglandins because it usually takes 48 to 72 hours for duct to close in a cyanotic CHD.

Pulmonary Atresia is associated with coronary sinusoids. Although none were present in our case, confirmed with CT pulmonary angiography.

Complete repair is always possible and results are good.

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