

A Case of ASD with Cyanosis in a Child

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Abstract: Cyanosis in child with ASD is an ominous sign. The common causes for cyanosis in a child with ASD includes TAPVC (total anomalous pulmonary venous connection), PAPVC (partial anomalous pulmonary venous drainage), persistent LSVC (left superior vena cava) with unroofed coronary sinus, Eisenmenger syndrome, Prominent Eustachian valve, common atrium and associated primary pulmonary hypertension.

Keywords: TAPVC, ASD, cyanosis, child, supra cardiac

1. Introduction

Cyanosis in a child with ASD warrants further evaluation. The common causes for cyanosis in a child with ASD includes TAPVC (total anomalous pulmonary venous connection), PAPVC (partial anomalous pulmonary venous drainage), persistent LSVC (left superior vena cava) with unroofed coronary sinus, Eisenmenger syndrome, Prominent Eustachian valve, common atrium and associated primary pulmonary hypertension. TAPVC (total anomalous pulmonary venous connection), PAPVC (partial anomalous pulmonary venous drainage), persistent LSVC (left superior vena cava) with unroofed coronary sinus, Eisenmenger syndrome, prominent Eustachian valve, common atrium and associated primary pulmonary hypertension. Here we report a case of ASD with cyanosis in a child which turned out to be a case of supra cardiac TAPVC.

2. Case Report

4 year old female child was diagnosed to have ASD and came to our institution for further management. Child gives history of recurrent respiratory tract infection and had poor weight gain. On examination, pulse – 110/mt, B p – 90/60 mm of Hg. Child had mild cyanosis and grade 1 clubbing. Oxygen saturation was 87% at room air. Examination of cardiovascular system revealed wide fixed splitting S2 with loud pulmonary component and an Ejection Systolic Murmur at pulmonary area. ECG showed in complete RBBB. Chest X- ray PA view showed figure of ‘8’ pattern with pulmonary plethora (fig 1). 2 D ECHO in apical 4 chamber view revealed large ASD, small LA and dilated RA and RV (fig 2). A common chamber (CC) was seen behind the LA. Suprasternal long axis view showed left vertical vein (LVV) draining in to right superior vena cava (RSVC) through innominate vein (INN) (fig 3). Patient was taken for cath angiography. Pulmonary Artery angiogram done in PA view in levophase showed right and left pulmonary veins draining in to common chamber and through left Vertical Vein, innominate vein and right superior vena cava draining in to RA (fig 4). Vertical Vein was selectively cannulated using 5 French Judkins Right Catheter and common chamber was entered. Hand injection of dye in to common chamber showed the venous conduit draining in to RA(fig

5). Oximetry data showed equal oxygen saturation in all four cardiac chambers. Patient later underwent successful corrective surgery.

3. Discussion

TAPVC defines the anomaly in which the pulmonary veins have no connection with the LA. Rather, pulmonary veins connect directly to one of the systemic veins or drain in to RA. The incidence of TAPVC is 0.008% of live births(1). It accounts for 2% to 3% of cases of congenital heart disease (2).

Based on the site of drainage of the pulmonary venous flow Darling and associates divided these anomalies into four subtypes: Type I, anomalous connection at the supra cardiac level; (45% of cases) Type II, anomalous connection at the cardiac level (26%); Type III, anomalous connection at the infra cardiac level (24%); and Type IV, mixed, with anomalous connections at two or more of the above levels (5%) (3). Male to female ratio is equal in TAPVC except for infra cardiac type, where it is 3 - 4:1. Developmentally TAPVC result from early atresia of the common pulmonary vein while pulmonary to systemic venous connections are still present(4).

The clinical manifestations depend on whether there is obstruction to the pulmonary venous connection. Although obstruction may occur with any anatomic type of TAPVC, the highest incidence is encountered with the infra cardiac type. If there is no obstruction, mild cyanosis, CHF and frequent pulmonary infection are the common manifestations. With obstruction, there is marked cyanosis, respiratory distress and pulmonary congestion. In TAPVC With obstruction age at death ranged from 2 days to 4.5 months (5). Without treatment 75 – 85% of TAPVC without obstruction will die in first year (6)

Medical management may be tried in patients with TAPVC without obstruction, in the form of diuretics, digoxin and correction of metabolic acidosis. Surgery is the definitive treatment. It is done emergent basis for obstructed TAPVC and within 6 months for non obstructed TAPVC.

4. Conclusion

Cyanosis in a child with ASD is an ominous sign and warrants further evaluation. The common causes for cyanosis in a child with ASD includes TAPVC, PAPVC, persistent LSVC with unroofed coronary sinus, Eisenmenger syndrome, prominent Eustachian valve, common atrium and associated primary pulmonary hypertension.

References

- [1] Correa-Villasenor A, Ferencz C, Boughman JA, Neill CA. Total anomalous pulmonary venous return: familial and environmental factors. The Baltimore-Washington Infant Study Group. *Teratology*. Oct 1991;44(4):415-28
- [2] Ferencz C, Rubin JD, McCarter RJ, et al. Congenital heart disease: prevalence at livebirth: The Baltimore-Washington Infant Study. *Am J Epidemiol* 1985;121:31-6.
- [3] Darling RC, Rothney WB, Craig JM. Total pulmonary venous drainage into the right side of the heart; report of 17 autopsied cases not associated with other major cardiovascular anomalies. *Lab Invest* 1957;6:44-64.
- [4] Edwards JE. Pathologic and developmental considerations in anomalous pulmonary venous connection. *Mayo Clin Proc* 1953;28:441-52.
- [5] Lucas RV Jr, Adam P Jr, Anderson RC. Total anomalous pulmonary venous connections to the portal venous system: a cause of pulmonary venous obstruction. *AJR Am J Roentgenol* 1961; 86: 561- 575.
- [6] Burroughs JT, Edwards JE. Total anomalous pulmonary venous connection. *Am Heart J* 1960; 59: 913 – 931.

Figure Title

Figure 1, Chest X- ray PA view showing figure of '8' pattern with pulmonary plethora

Figure 2- Apical 4 chamber view showing large ASD, small LA and dilated RA and RV . A common chamber (CC) was seen behind the LA

Figure 3- Suprasternal view showing left vertical vein (LVV) draining in to right superior vena cava (RSVC) through innominate vein (INN)

Figure 4 -Pulmonary Artery angiogram in levophase showing left Vertical Vein , innominate vein and right superior vena cava.

Figure 5 -Hand injection of dye in to common chamber showing the venous conduit draining in to RA.



Figure 1: Chest X- ray PA view showing figure of '8' pattern with pulmonary plethora

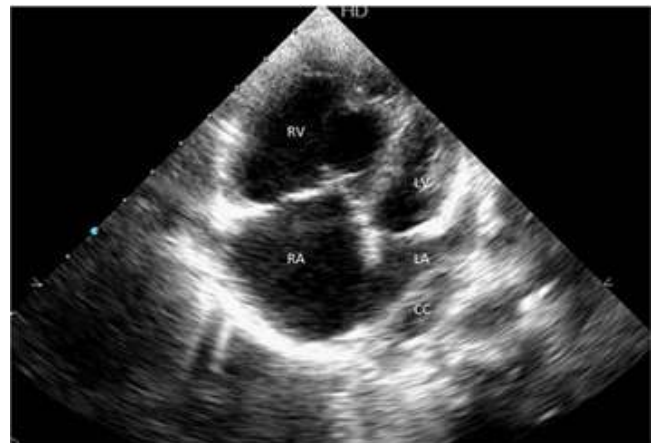


Figure 2: Apical 4 chamber view showing large ASD, small LA and dilated RA and RV . A common chamber (CC) was seen behind the LA

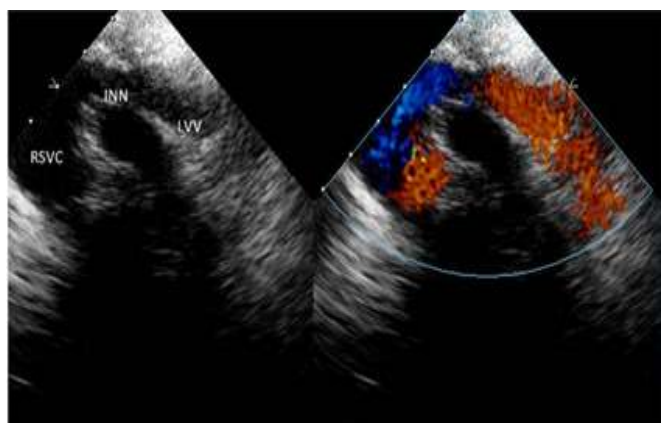


Figure 3: Suprasternal view showing left vertical vein (LVV) draining in to right superior vena cava (RSVC) through innominate vein (INN)

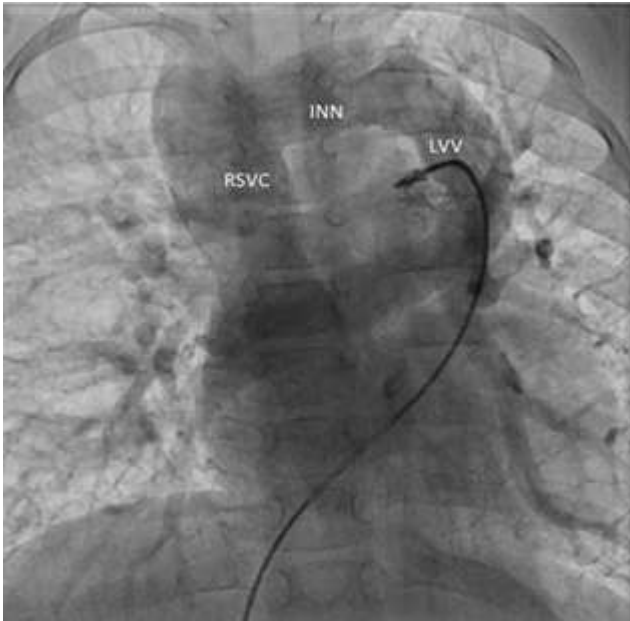


Figure 4: Pulmonary Artery angiogram in levophase showing left Vertical Vein, innominate vein and right superior vena cava

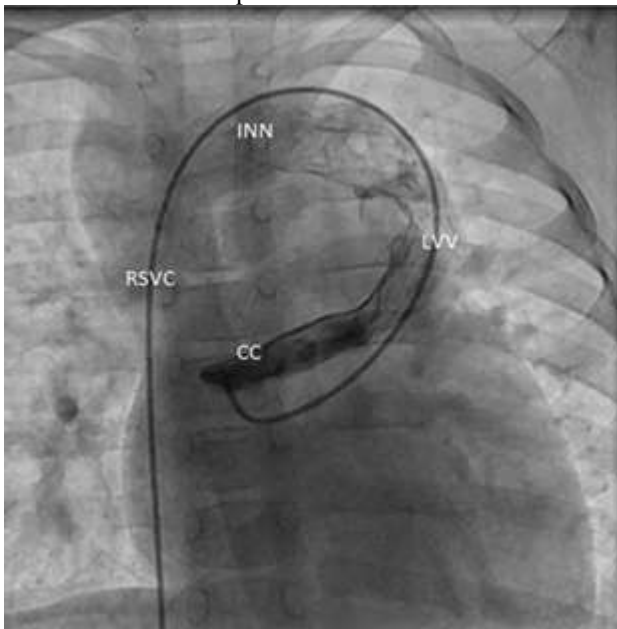


Figure 5: Hand injection of dye into common chamber showing the venous conduit draining into RA