Cyanotic atrial septal defect: Literature review

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Abstract
Atrial septal defects (ASD) constitute a tenth of all congenital heart disease in children. It is a common cause of acyanotic congenital heart defects in children. Cyanosis in a case of ASD is something which should hint that the condition is not a simple ASD and should prompt to search for causes for the same. The conditions of cyanosis with ASD have been discussed in this article.

Keywords: Atrial Septal Defect (ASD), Congenital Heart disease (CHD), Cyanosis.

The incidence of Congenital heart diseases in our country is estimated to be 8-10 per 1000 live births, out of which ASDs have an incidence of 0.56 per 1000 live births.[1] Isolated ASD is considered the most common CHD in adults presently.[2] On the basis of location, ASDs are of four types with secundum ASD (75%) being the most common followed by primum ASD (20%), sinus venosus ASD (5%) and coronary sinus ASD.[3] The female to male ratio for secundum ASD is 2:1 but is equal for sinus venosus ASD.[4,5] Mostly these ASDs remain asymptomatic during early years of life and generally become symptomatic in the 3rd or 4th decade of life with presenting symptoms being dyspnoea, fatigue, palpitations, atrial flutter and fibrillation.

This is basically an acyanotic congenital disease with communication between two low pressure chambers of the heart. Flow across the ASD depends on the size or relative compliance of the ventricles.[2] Cyanosis in a case of ASD should make our minds to search for the additional lesion or cause which is causing this acyanotic condition to behave as a cyanotic condition.

The causes of cyanosis in a case of ASD can be due to the following conditions:

1. Eisenmenger's syndrome with elevated pulmonary arterial pressure and pulmonary vascular resistance
2. Anomalies in systemic venous drainage including anomalies of connection, straddling SVC or IVC in SV ASD or abnormal routing of IVC blood due to a prominent Eustachian valve
3. Elevated right atrial pressures as in right atrial myxoma, right ventricular infarction, mechanical ventilation with raised PEEP
4. Platypnea- orthodeoxia syndrome.[2]

With elevation of PA pressures and further PVR, the compliance of the right ventricle changes and with further time, the reversal of shunt occurs and leads to eisenmengerization.

The systemic venous blood if it drains to the left atrium, with lead to cyanosis. This will be an obligatory shunt and will account for constant desaturation in these children. The most common form among these is the persistent left SVC draining into left atrium through unroofed coronary sinus. Sometimes, SVC and/ or IVC too connect anomalously.[6] A few case reports of totally anomalous systemic venous connections are also available.[7] Sometimes the valve of IVC i.e. the Eustachian valve is prominent which can lead to selective streaming of systemic IVC blood to left atrium leading to cyanosis. A few cases are also reported in which the regurgitant jet of tricuspid valve is directed towards the ASD in turn leading to the systemic blood of right ventricle to enter into left atrium and hence the cyanosis.

Elevated right atrial pressures due to any cause will result in abnormal right ventricle filling. In case of right atrial myxomas, there is mechanical obstruction to tricuspid valve flow and this will lead to decreased right ventricle filling. Cases in which there is reduced right ventricle systolic function like myocardial infarction can lead to functional tricuspid valve inflow restriction and reversal of shunt across ASD and hence cyanosis.[2] In patients who are on mechanical ventilator, if PEEP is high, it results in right to left shunting across the ASD and so cyanosis.

Platypnea- orthodeoxia syndrome is an underdiagnosed condition in which there is desaturation and cyanosis along with dyspnoea when changing from recumbent to an upright position. The cause has been attributed to the right to left shunting which occurs across the ASD on assuming a standing position as the orientation of the inter- atrial septum changes.[8]

So, cyanosis in a case of ASD should prompt us to take the condition seriously and hunt for the cause in addition to this shunt alone, so that appropriate management can be undertaken.

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