Cyanosis in a patient with an atrial septal defect (ASD) is uncommon, albeit an important clinical sign. It can result with the development of right-to-left (R-L) shunt across an ASD initially shunting left to right (L-R) in view of elevated pulmonary artery (PA) pressures, as in Eisenmenger’s syndrome. However, cyanosis may present early in a subset of patients having normal PA pressure due to the presence of associated anomalies which favor R-L shunting across an ASD. We present a case of a cyanotic child with an echocardiographic diagnosis of ostium secundum ASD with severe tricuspid regurgitation (TR).

A 5-year-old male child weighing 13.5 kg presented to our hospital with chief complaints of fast breathing and bluish discoloration of the body for 1 year of age. The physical examination revealed clubbing, cyanosis, and raised jugular venous pressure. A Grade III pansystolic murmur was heard at lower sternal border, and soft non-tender hepatomegaly was evident on per-abdominal examination. The electrocardiogram demonstrated right axis deviation and the chest X-ray was unremarkable with normal pulmonary bronchovascular markings. Two-dimensional transthoracic echocardigraphy demonstrated situs solitus, levocardia, atrioventricular, and ventriculoarterial concordance with a large Osteum secondum-ASD (7 mm) shunting R-L and severe low-pressure TR.

Anesthesia was induced and maintained as per standard institutional protocols. Transesophageal echocardiography (TEE) was performed with S7-3t Philips ultrasound probe and machine (Philips iE33 model, Bothell, WA, USA). A large OS-ASD (1.3 cm) with R-L shunt and enlargement of the right heart chambers (right atrial [RA]/right ventricle) was the most obvious finding. The surgeon was requested to assist in needle transduction of the pulmonary pressures, wherein the PA pressures were well within normal range. Thereby, the clinical scenario constituted an OS-ASD shunting R-L despite normal PA pressures [Figure 1].

Subsequently, the TEE examination revealed significant TR. In the mid-esophageal 4 chamber view, the tricuspid valve (TV) appeared dysplastic with annular diameter of 2.98 cm (BSA 0.56 m²). The anterior tricuspid leaflet (ATL) was prolapsed with the complete absence of chordae tendineae to support the ATL. This resulted in severe low-pressure TR (PG 25 mmHg) with vena contracta of 1.36 cm with abnormal streaming of systemic venous blood into the LA. There were no echocardiographic signs of a dilated coronary sinus or any abnormal venous connection.

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Surgical exploration confirmed the presence of large OS-ASD, dysplastic TV, and absence of chordae tendineae to support the ATL. The surgeon decided to repair the TV with alferri stitch and close the OS-ASD. The patient was shifted to Intensive Care Unit (ICU) on minimal inotropes (injection Dopamine 5 μg/kg/min and injection Adrenaline 0.1 μg/kg/min), which got tapered in ICU and patient was successfully extubated on 1st postoperative day. The patient was discharged from the hospital after 5 days.

An ASD typically shunts L-R in the absence of pulmonary hypertension.[1] Certain meticulously carried out pressure-flow studies have demonstrated a small R-L shunt in simple ASDs occurring during brief periods of a positive pressure gradient from R-L during the cardiac cycle. This is believed to occur either in early atrial systole or during early ventricular systole.[2,3] Nevertheless, a clinically significant R-L shunt across an ASD results in the presence of certain anatomical and physiological peculiarities. Table 1 summarizes the important causes of reversal of shunt in an ASD.

In the index case, ASD was associated with a congenital TR leading to the development of R-L shunt across the defect. The mechanism of TR was discovered as the prolapse of ATL with the abnormal streaming of systemic venous blood into the LA and resultant cyanosis. Tanji et al. also reported a case of a 5-year-old girl with ASD of the secundum type with congenital TR which was associated with peripheral pulmonary stenosis.[4] Studies have revealed TR as an independent determinant of the R-L shunt in ASD even in the absence of the reversal of pressure gradient between the L-R atrium.[5]

TR is frequently associated with an ASD, which largely remains functional in origin.[6] RVSP (calculated as RA pressure + 4 [vTR]²) in such a case with TR secondary to pulmonary hypertension gives an estimate of increased pulmonary artery systolic pressure. The morphology of the tricuspid valvular leaflets remains largely normal except annular dilatation in the face of elevated right-sided pressures. However, the co-existence of an ASD and TR in the present case could not be attributed to pulmonary hypertension alone.

To conclude, cyanosis may manifest in patients of ASD despite normal PA pressures with potentially treatable conditions. The authors wish to elucidate the fact that a relatively early onset of cyanosis in ASD, where the physical examination, electrocardiogram and chest roentgenogram fail to suggest elevated PA pressures, one might be dealing with an altered physiology across the ASD, the cause for which needs to be discovered for appropriate etiology directed management. This highlights the importance of a meticulous TEE evaluation of the TV morphology in addition to assessing the gradients across the valve in a patient with such a clinical background.

**Table 1:** Summarizes the important causes of reversal of shunt in an atrial septal defect

| Significant R-L shunt across an ASD |
|-------------------------------------|
| Elevated PA pressures and PVR |
| Eisenmenger’s syndrome |
| Normal PA pressures |
| Systemic venous blood drainage anomalies-persistent LSVC draining in LA or unroofed CS, SVC/IVC straddling or abnormal streaming of IVC blood or TR jet |
| Abnormalities of RV filling leading to high RAP-RA myxoma, RV infarction, diastolic dysfunction or cardiomyopathy, PPV with high PEEP |
| Platynea-orthodeoxia syndrome in patients with coexisting aortic aneurysm, loculated pericardial effusion or postlobectomy |

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**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.
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