Case report

Right sided endocarditis and right heart failure in a child with ventricular septal defect – A case report

Abstract:

Background:

Right-sided endocarditis, though less common than its left-sided counterpart, remains a significant concern, particularly in the context of congenital heart defects such as ventricular septal defects (VSDs). In some instances, the narrowing of ventricular septal defects by the tricuspid leaflet can lead to organic tricuspid regurgitation (TR), predisposing patients to recurrent right heart failure.

Case presentation:

We present a rare case of a 10-year-old girl experiencing recurrent right heart failure secondary to extensive tricuspid regurgitation associated with a ventricular septal defect. Over two years, her symptoms progressed from New York Heart Association (NYHA) class II to IV, with accompanying signs of anasarca, respiratory distress, and poor feeding. Despite previous episodes of infective endocarditis and recommended surgical intervention, familial consent was initially lacking, leading to deferred surgery. Upon presentation to our centre, the patient was diagnosed with severe tricuspid regurgitation and a restrictive ventricular septal defect, necessitating urgent surgical correction. However, post-surgery, she developed complete heart block and subsequent complications, culminating in her demise within 24 hours.

Conclusions:

This case underscores the potential complications of small congenital heart defects, particularly when complicated by infective endocarditis. The occurrence of sub clinically massive TR

resulting in recurrent right heart failure in a paediatric patient highlights the need for vigilance in monitoring and timely intervention. Early recognition and appropriate management are essential in preventing devastating complications associated with infective endocarditis in the setting of congenital heart disease.

Key words:

Ventricular septal defect, right heart failure, right sided endocarditis, tricuspid regurgitation

Introduction:

Right-sided endocarditis is a recognized complication of small ventricular septal defects (VSDs). Organic tricuspid regurgitation (TR) can arise as a consequence of VSD-induced right-sided infective endocarditis (IE). However, the occurrence of sub clinically massive, torrential TR resulting in recurrent heart failure in a paediatric patient is scarcely documented. We report a case of recurrent right heart failure attributed to significant tricuspid regurgitation associated with a ventricular septal defect in a child.

Case Presentation:

A 10-year-old girl presented to our centre with progressively worsening dyspnoea on exertion, worsened from NYHA class II to class IV over a span of two years. Additionally, she exhibited anasarca, initially manifesting in the lower limbs and subsequently spreading throughout her body, alongside symptoms of easy fatigability, respiratory distress, and poor feeding.

Physical Examination:

Clinical examination revealed dental caries in multiple tooth, grossly elevated jugular venous pressure with prominent CV waves, anasarca, and hepatomegaly. Auscultation identified soft S1, loud P2, and a pan-systolic murmur graded V/VI in the left parasternal area, accentuated during inspiration without radiation. Bilateral basal crepitations were noted, in addition to firm hepatomegaly measuring 13 cm in span.

Past History:

The patient was diagnosed with a VSD at the age of 2 years and was advised regular medical follow-up due to anticipated spontaneous closure. However, at ages 7, 8, and 9 years, she experienced episodes of infective endocarditis with organisms Staphylococcal aureus, Streptococcus sinensis with resultant right heart failure, necessitating decongestive measures and antibiotic therapy at an outside hospital. Upon presentation to our centre at 9 years of age, she was diagnosed with a restrictive VSD accompanied by severe TR. Despite a recommendation for surgical intervention, familial consent was lacking, leading to deferred surgery. Three months later, she presented with acute decompensated right heart failure, necessitating multiple inotropic agents, decongestive measures, and intravenous antibiotics. Subsequent evaluation revealed Staphylococcal aureus septicaemia without intracardiac vegetations, prompting treatment as infective endocarditis in the context of recurrent episodes and decompensated heart failure.

Investigations:

Laboratory investigations including hemogram, liver function tests, renal function tests, and urine analysis returned within normal limits. Primary and secondary immunodeficiency work-up, as well as viral markers, were negative. Radiographic evaluation via chest X-ray demonstrated cardiomegaly with a cardiothoracic ratio of 0.90 (Figure 01) and signs of pulmonary venous congestion. Electrocardiography (ECG) revealed sinus rhythm, right atrial (RA), right ventricular (RV) enlargement, and P pulmonale. Two-dimensional echocardiography (2D ECHO) unveiled grossly dilated RA with RA area of 31.6cm2 and RV, restrictive VSD with a size of 4.9mm with left-to-right shunt with a pressure gradient of 77mm Hg (Figure 02A, 02B, 02D) (Video 01), deformed tricuspid leaflets, tricuspid annular diameter of 34mm (Figure 02C), massive central tricuspid regurgitation (Video 02) with moderate pulmonary artery hypertension (PAH) (Right Ventricular Systolic Pressure= 46mm Hg+ Right Atrial Pressure) (Figure 02E), moderate pericardial effusion more on the anterior side, and dilated inferior vena cava (IVC). Cardiac computerized tomography revealed similar findings (figure 03) with mild pulmonary venous congestion without evidence of tricuspid, pulmonary or branch pulmonary artery vegetations, consolidation or collapse.

Management:

Given the high-risk nature of the case, surgical closure of the VSD coupled with tricuspid valve repair under cardiopulmonary bypass was deemed appropriate due to marked right heart dilatation and torrential TR. VSD closure was achieved utilizing a pericardial patch secured with pledgeted interrupted sutures, alongside insertion of a 26 mm rigid tricuspid annuloplasty ring, resulting in trivial TR and closure of the VSD. However, the patient experienced complete heart block, low cardiac output syndrome within six hours post-surgery, precipitating severe RV dysfunction leading to anuric renal failure requiring peritoneal dialysis and eventual demise within 24 hours.

Discussion:

While less prevalent than left-sided endocarditis, right-sided endocarditis carries its own clinical significance. Native valve endocarditis typically arises secondary to identifiable risk factors, with congenital heart defects, notably ventricular septal defects, representing a common predisposing condition. The mechanism involves constriction and diminution of the VSD size by the septal tricuspid leaflet, aortic cusp prolapse, or muscle overgrowth, rendering them susceptible to infective endocarditis. The increased pressure and trauma to the septal tricuspid leaflet make it more susceptible to infective endocarditis. Vegetation formation can lead to the destruction of the leaflet, resulting in tricuspid regurgitation. Tricuspid regurgitation may occur due to various mechanisms, including restriction caused by a septal aneurysm, distortion of the septal leaflet by a high-velocity jet, and the Gerbode effect, which can mimic tricuspid regurgitation[1,2,3].Closure of the ventricular septal defect can alleviate these issues with tricuspid valve regurgitation[4,5]. While closure of the VSD can resolve these regurgitations of the tricuspid valve, complete leaflet destruction causing chronic volume overload and further aggravating TR is less common (Figure 04). Chronic subclinical volume overload with recurrent right heart failure secondary to infective endocarditis is extremely rare, especially in the paediatric population. Most cases respond well to antibiotic therapy, but approximately 20% require surgery, especially in cases of intractable refractory right heart failure, large vegetations, uncontrolled septicaemia, or vegetations with complications such as systemic embolization or damage to surrounding structures[6]. The timing of surgery is debatable in VSD with TR, with some authors recommending surgery after the control of active infection to avoid patch infection[7]. Tricuspid valve reconstruction requires extensive and aggressive debridement, valve reconstruction avoiding artificial material, and using as little foreign material as possible. Congenital heart disease with IE had 11% mortality during the active phase of infection and

8.0% in the medically managed group[8]. The cause of mortality was surgery, renal failure, and cardiac failure as in our case[8].

Conclusions:

Infective endocarditis should be suspected in cases of prolonged fever or acute decompensated state of congenital heart disease. Always look for complications in case of unusual presentation of small heart defects. Timely recognition and prompt management can prevent devastating complications in infective endocarditis.

Consent & Ethical approval

Informed consent was obtained from the parents of the child for the publication. The identity of the patient has not been divulged in any form. Institutional ethics approval is not required.

List of abbreviations:

VSD: Ventricular septal defect

TR: Tricuspid regurgitation

IE: Infective endocarditis

NYHA: New York Heart association

ECG: Electrocardiography

RA: Right atrium

RV: Right ventricle

2D ECHO: 2-dimensional echocardiography

PAH: Pulmonary artery hypertension

IVC: Inferior vena cava

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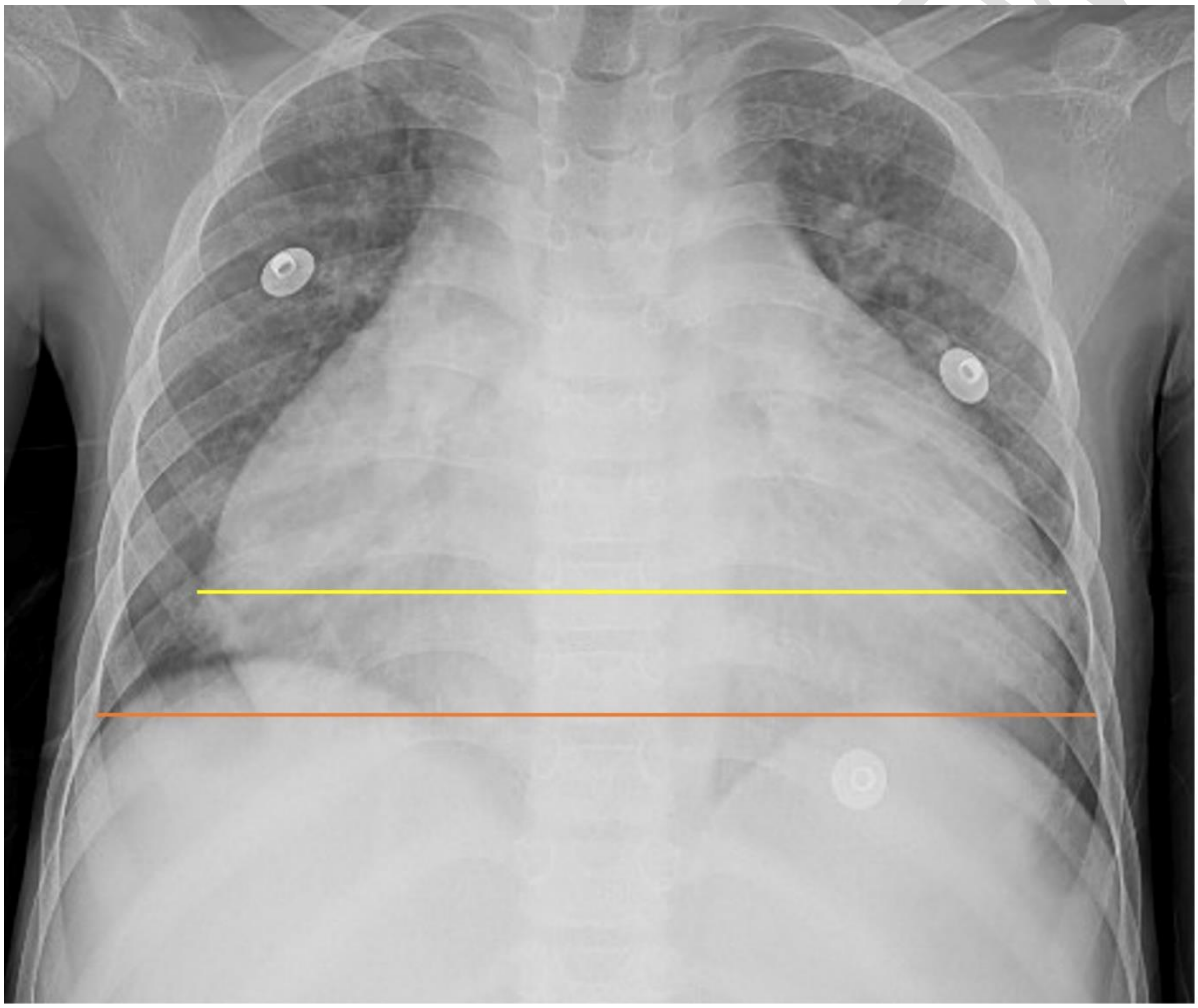

Figure 01: Chest X ray showing cardiothoracic ratio of 0.90, yellow line cardiac diameter 0.9cm, red line thoracic diameter of 1.0cm

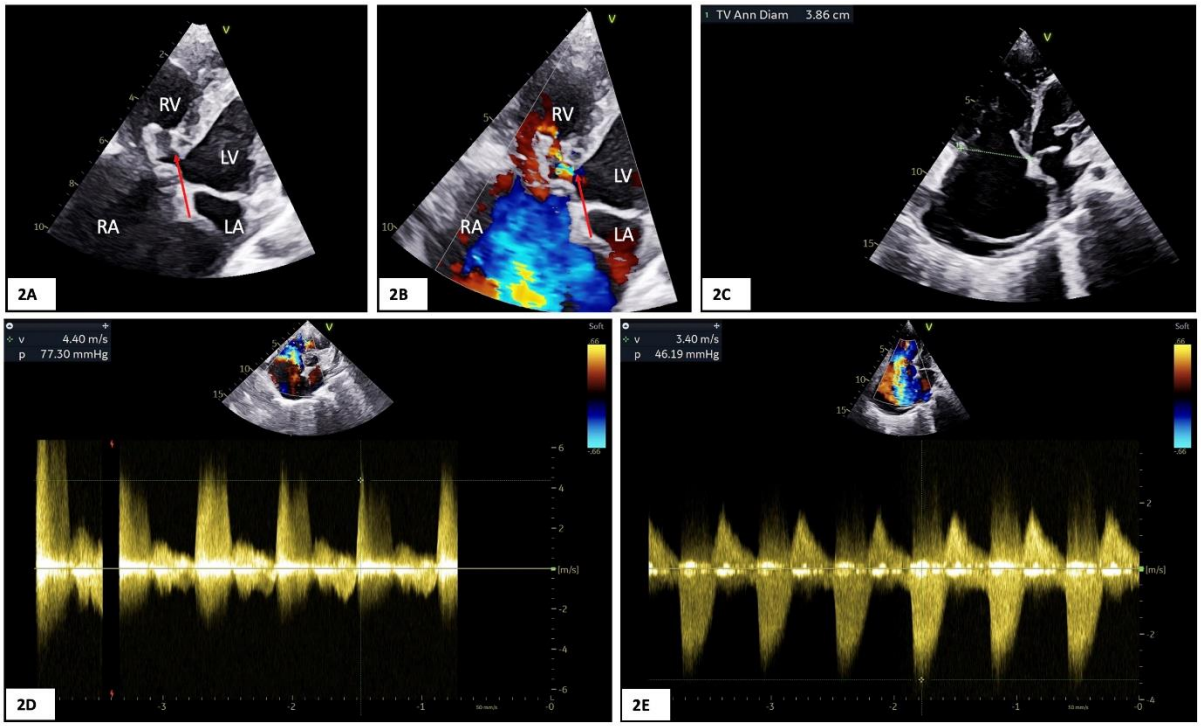

Figure 02: Two-dimensional echocardiography. 2A, 2B: Apical four chamber view without and with color flow image demonstrating ventricular septal defect indicated by red arrow. 2C: Apical four chamber view showing tricuspid annular diameter of 3.86cm and moderate pericardial effusion behind right atrium. 2D: Continuous wave doppler across the ventricular septal defect showing pressure gradient of 77mm Hg. 2E: Continuous wave doppler across the tricuspid regurgitation showing pressure gradient of 46mm Hg.

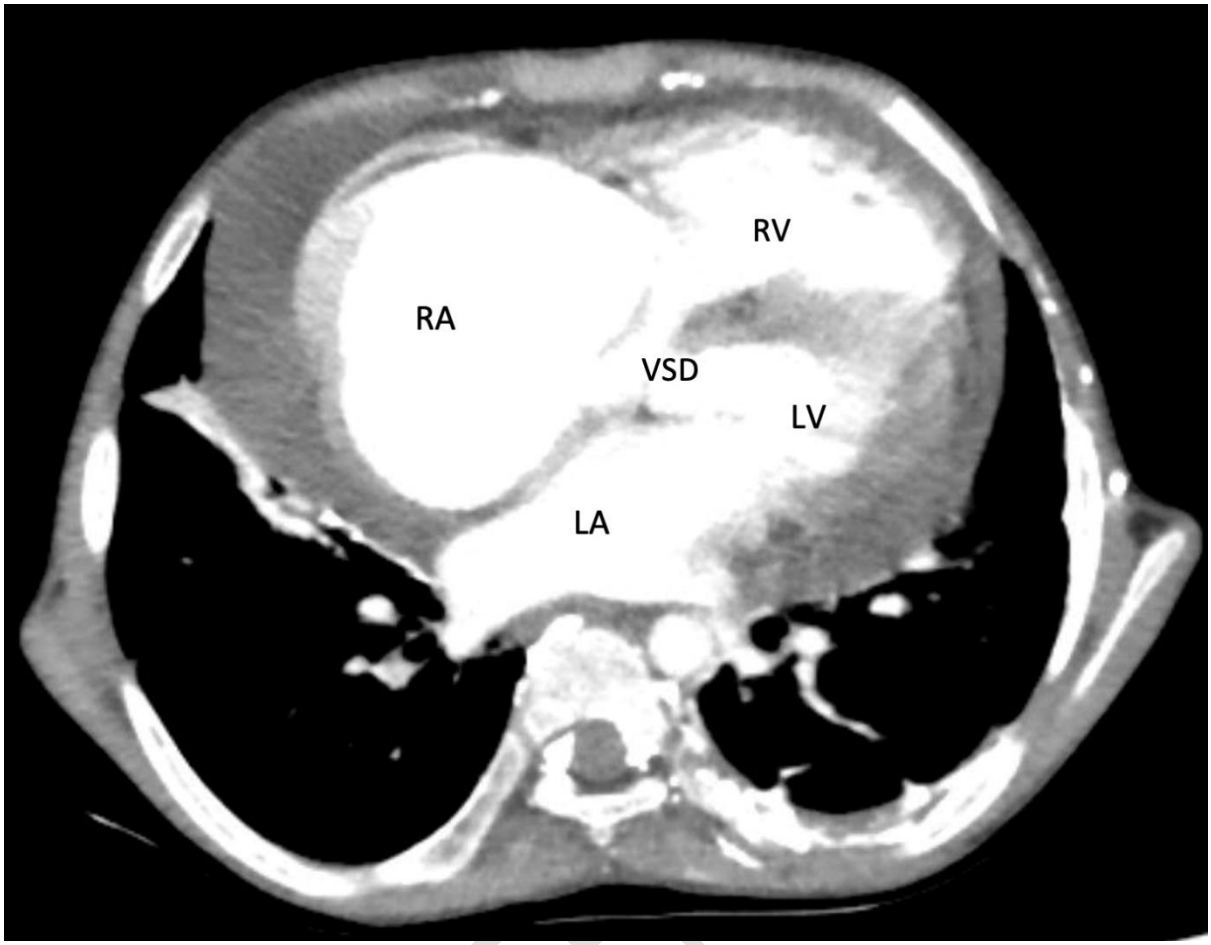

Figure 03: Cardiac Computerized tomography contrast image demonstrating ventricular septal defect in five chamber view.

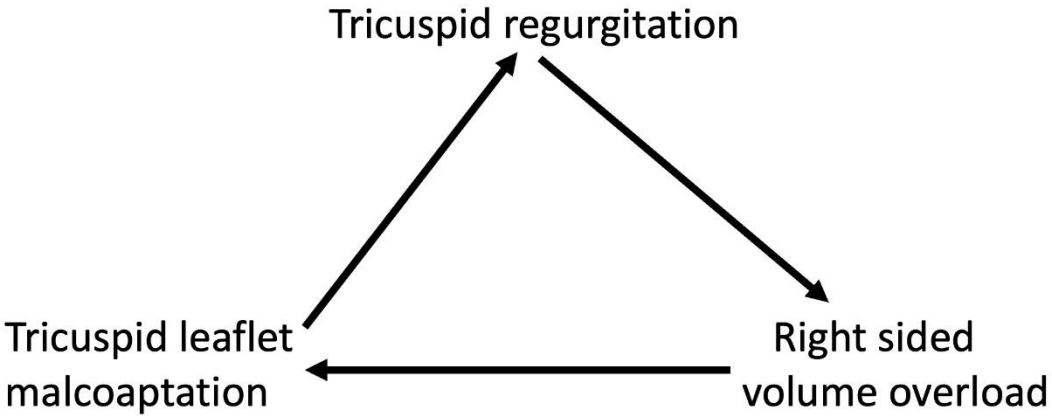

Figure 04: Flow diagram depicting mechanism of tricuspid regurgitation.

Supplementary video 01: Modified Apical four chamber view demonstrating severe tricuspid regurgitation and ventricular septal defect with left to right shunt.

Video 1.mp4

Supplementary video 02: Apical four chamber view demonstrating severe tricuspid regurgitation.

Video 2.mp4