

Isolated Prolapse of the Tricuspid Valve with Severe Tricuspid Regurgitation in a Young Adult

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Abstract

We present a case report of a young male adult with severe tricuspid regurgitation due to isolated prolapse of the tricuspid valve. The prolapse was caused by a congenital abnormality of the valvular and subvalvular apparatus of the tricuspid valve.

Keywords: Isolated prolapse; Tricuspid regurgitation; Tricuspid valve

Case Report

A 23-year-old man presented with symptoms of palpitations during exercise and mild reduced exercise tolerance. On examination a pansystolic murmur was audible at the left lower sternal edge. His medical history revealed a heart murmur when he was about ten years old, but was not further investigated. Further on he had no medical history. The electrocardiogram showed a sinus rhythm with partial right bundle branch block, the duration of the QRS complex being 112 milliseconds. There were no rhythm abnormalities during the exercise test nor with holter monitoring. Echocardiography showed mild dilatation of the right ventricle and the right atrium with a prolapse of the anterior leaflet of the tricuspid valve (figure 1).

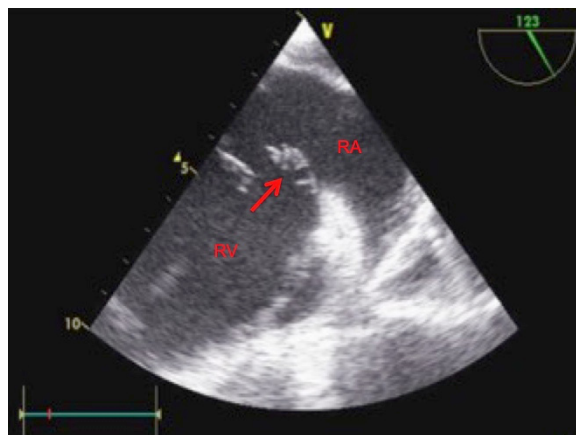


Figure 1: Transesophageal echocardiography: prolapse of the anterior leaflet of the tricuspid valve (red arrow) RA= right atrium, RV= right ventricle

Secondary to the prolapse there was a severe eccentric TR (figure 2): Proximal Isovelocity Surface Area (PISA) radius of 9mm, Effective Regurgitant Orifice Area (EROA) of 70 mm² and a regurgitant volume of 48 mL. There were no signs of pulmonary hypertension (tricuspid regurgitation velocity 2,6 m/s), nor were there any structural or functional abnormalities of the mitral valve.

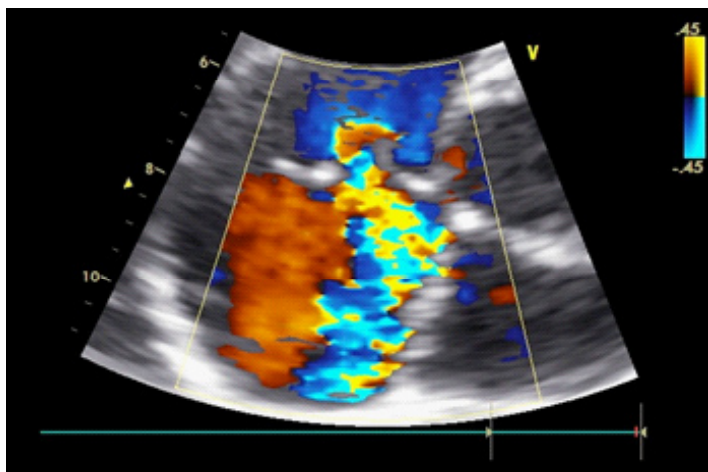


Figure 2: Transthoracic echocardiography: severe tricuspid regurgitation (blue colour)

The patient was referred to the cardiothoracic surgery department. Minimally invasive heart surgery was performed. Peri-operative a prolapse of the anterior leaflet of the tricuspid valve was confirmed (figure 3). There were no chordae attached to the anterior portion of the prolapsed leaflet, the anterior papillary muscle was absent. All chords originated from a small septal and a big trifid posterior papillary muscle. In addition, a moderate dilatation of the tricuspid annulus was noticed. Tricuspid valve repair was performed with Gore-Tex neo-chordae and annuloplasty by the insertion of a Carpentier Edwards 30 mm ring (figure 4). Post-operative there was no TR anymore.

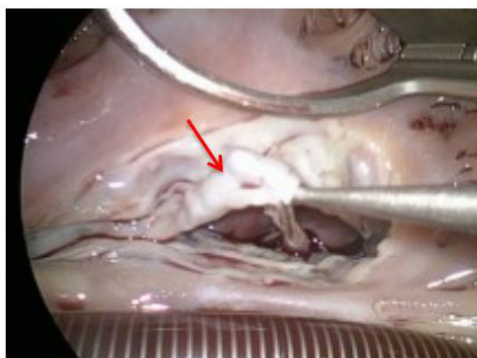


Figure 3: Peri-operative: prolapse anterior leaflet of the tricuspid valve (red arrow)

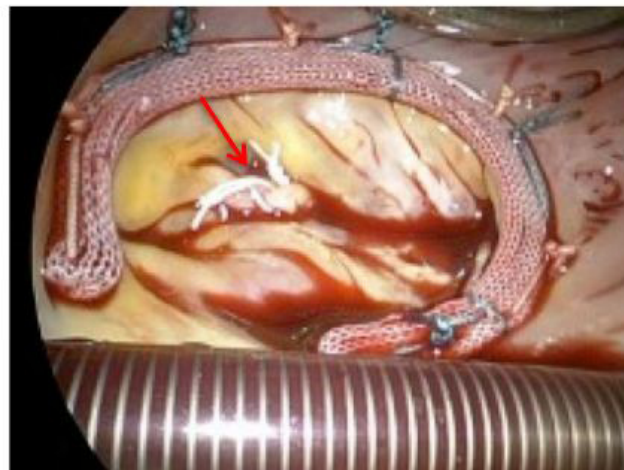


Figure 4: Post-operative: suture of the neo-chordae (red arrow) to the anterior tricuspid leaflet and annuloplasty with a Carpentier Edwards 30mm ring

Further follow-up after one and two year showed a good result of the valvuloplasty without any regurgitation of tricuspid valve.

Discussion

The term isolated prolapse of the tricuspid valve refers to prolapse involving only the tricuspid valve. Tricuspid valve prolapse has been found more frequently to be associated with mitral valve prolapse or with other cardiac and lung diseases. The current literature of isolated prolapse of the tricuspid valve is limited to a small number of reports. At this moment at least 40 cases of isolated prolapse of the tricuspid valve have been reported. In 1985 Weinreich et al. documented 34 cases. These report cases were all in association with left ventricular abnormalities, right ventricular pressure overload or direct valvular injury [3]. In addition, Weinreich et al. described three new cases of isolated prolapse of the tricuspid valve [4], these cases were associated with dilated ischemic cardiomyopathy, hypertensive heart disease and chronic lung disease. In 1990 Mittal et al. reported a severe TR due to isolated tricuspid valve prolapse [5]. Tricuspid valve prolapse is rarely found as an isolated finding. It appears to be a relatively unknown anatomico-clinical entity although it is of clinical importance since this condition may be associated with significant TR, cardiac arrhythmias and bacterial endocarditis. In isolated TR, the right ventricle progresses to a volume-overloaded state, and concurrently, both right atrial dilation and right atrial pressure elevation take place. With long-standing significant TR, patients develop signs of right heart failure (ascites, peripheral oedema, weight gain, hepatic dysfunction). Two-dimensional (2D) transthoracic echocardiography remains the cornerstone in establishing the diagnosis of TR and grading its severity by providing

qualitative, semi-quantitative and quantitative parameters. Isolated TR is associated with increased mortality [6].

In our case report the prolapse of the anterior tricuspid leaflet was caused by a congenital abnormality of the valvular and sub-valvular apparatus of the tricuspid valve. TR secondary to congenital anomalies is rare in the absence of Ebstein's malformation. It may be due to abnormal development of the chords, papillary muscles or the valvular leaflets. Severe TR is often well tolerated in early childhood, although long-standing and progressive volume loading of the right heart leads to symptoms of decreased exercise tolerance and may predispose to arrhythmias in the long term. Abrams et al. reported in 2005 three cases of young patients (8-, 11- and 12 years-old) with severe TR related to anomalies of the chords supporting the anterior leaflet of tricuspid valve [7]. Shortened chords leading to tethering of the leaflet were seen in two cases and hypoplasia of the leaflet in the other. One case of an 11-year-old boy with an isolated prolapse of the tricuspid septal leaflet was described by Patanè et al [8]. in 2005. Due to the small number of case reports, there are no clear guidelines dictating the need for surgical intervention. The onset of symptoms and/or evidence of reduced exercise capacity or arrhythmias are a good indication for surgery. When surgery is performed, valve repair is preferred over replacement. Valve replacement is associated with a higher mortality rate [9]. Surgical repair [10] is possible in the vast majority of cases of tricuspid valve and chordal anomalies. In the setting of absent or shortened chords, implantation or chordal lengthening using polytetrafluoroethylene, coupled with annuloplasty and commissural valvoplasty, has produced excellent results in the immediate and medium term follow-up.

Conclusion

Isolated prolapse of the tricuspid valve is a rare disease. When significant TR develops, this condition can be associated with cardiac arrhythmias and possibly bacterial endocarditis. Symptomatic severe TR in patients with isolated prolapse of the tricuspid valve can be treated by surgical valve repair.

Conflict of Interest

None declared

References

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