

Congenital cleft of right atrioventricular valve in partial atrioventricular canal defect: a rare entity

Trushar Gajjar, Neelam Desai

Received: January 14, 2016 Accepted: March 17, 2016 Published online: July 21, 2016

ABSTRACT

A cleft of the right atrioventricular valve is an extremely rare congenital entity. Herein, we report a case of 15 year old female diagnosed with primum atrial septal defect with cleft in both left and right atrioventricular valves. This diagnosis was obtained using an echocardiography. Surgical closure of the clefts of the left as well as the right atrioventricular valves was completed with a closure of the primum atrial septal defect. We will discuss the morphology, diagnostic modalities and management guidelines for this rare entity.

Keywords: Cleft right atrioventricular valve; echocardiography; partial atrioventricular canal defect; right atrioventricular valve insufficiency.

A congenital cleft of the right atrioventricular (AV) valve is very rare. It may or may not be associated with other cardiac anomalies.^[1-3] The most commonly associated congenital cardiac anomalies are atrial septal defect (ASD), ventricular septal defect, and tetralogy of Fallot.^[1-3] In this article, we report a case diagnosed partial atrioventricular canal defect, moderate left and right atrioventricular valve regurgitation. Diagnosis was obtained with echocardiography and surgical closure of the cleft in left and right atrioventricular valve was done with closure of primum ASD. We discuss here morphology, diagnostic modalities and management guidelines for this rare entity.

CASE REPORT

A 15-year-old female was referred to our outpatient department with a history of dyspnea New York Heart Association (NYHA) class II and a recurrent respiratory tract infection. Upon physical examination, there was a grade 2/6 pansystolic murmur at the apex and the subxiphoid area. The second heart sound was loud with wide and fixed split. The chest X-ray showed a cardiothoracic ratio of 0.6, with an enlarged right atrium and main pulmonary artery. The electrocardiogram showed deviation of the sinus rhythm in the left axis. The two dimensional echocardiography revealed: situs solitus, levocardia, normal pulmonary venous drainage and interrupted inferior vena cava (IVC) with hemiazygos continuation. It also showed dilated coronary sinus, ostium primum and an ostium secundum defect. Furthermore, they

discovered a left to right shunt, a cleft in the anterior left AV leaflet with mild and moderate regurgitation, a moderate right AV valve regurgitation and a cleft in the right AV valve. (Figure 1).

The right ventricular systolic pressure (RVSP) was 35 mmHg, showing normal biventricular function. The patient was taken up to surgery and the intraoperative findings showed the cardiomegaly in the left superior vena cava was draining into the dilated coronary sinus. Large primum ASD, complete cleft in left AV valve leaflet with moderate regurgitation, complete cleft in right AV valve with moderate regurgitation. The cleft in the left AV valve was sutured with interrupted 5-0 polypropylene sutures. The primum ASD was closed with an autologous untreated pericardial patch which keeping the coronary sinus on the right atrial side. The cleft in the right AV valve was repaired with 5-0 polypropylene suture (Figure 2). The posterior right AV valve annuloplasty was done using 4-0 polypropylene pledgeted suture.

The intraoperative transesophageal echocardiography revealed no residual shunt in the trivial left and right AV valve regurgitation and good biventricular function. The postoperative course

Sri Sathya Sai Institute of Higher Medical Sciences-Prasanthigram, CTVS Department, Andhra Pradesh, India.

Corresponding author: Trushar Gajjar, MD. Sri Sathya Sai Institute of Higher Medical Sciences - Prasanthigram, CTVS Department, Andhra Pradesh, India.
Tel: +919700355411 e-mail: trushargajjar@gmail.com

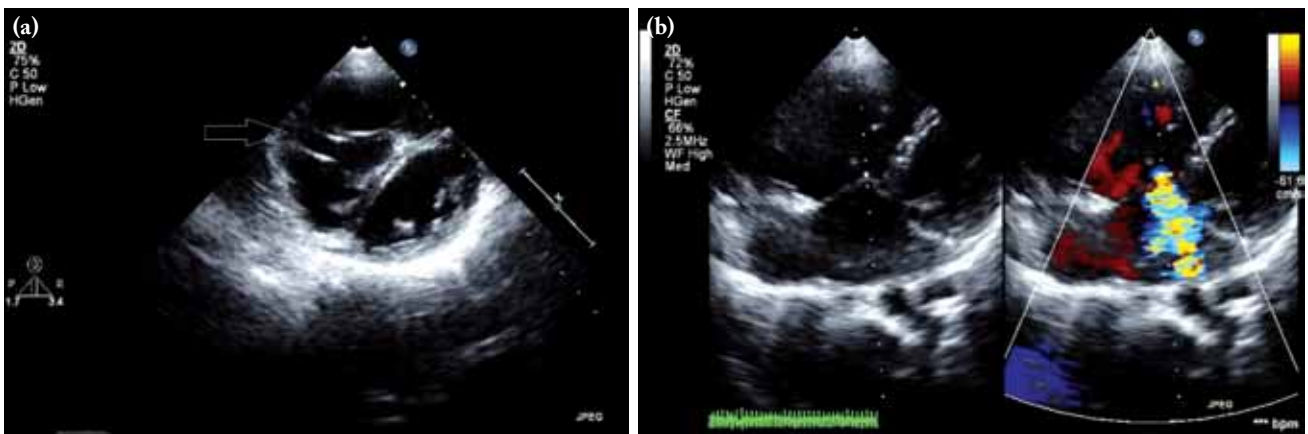


Figure 1. Transthoracic echocardiography. (a) Parasternal short axis view showing, a cleft in the right atrioventricular valve (Arrow). (b) Apical four chamber view showing a well coapted right atrioventricular valve with moderate regurgitation.

was uneventful. The transthoracic echocardiography before discharge showed, there was not any residual ASD, trivial left and right AV valve regurgitation, RVSP 22 mmHg, mild right ventricular dysfunction, no pericardial effusion, and normal left ventricular function.

During the three-month follow-up appointment, the patient was in NYHA class I, and the echocardiography showed, no residual shunt or trivial left and right AV valve regurgitation. There was good ventricular function. A written informed consent was obtained from the patient.

DISCUSSION

The cleft of the left AV valve is a well known entity and it is most commonly associated with atrial

and ventricular septal defects, endocardial cushion defects, transposition of the great arteries and even as isolated anomalies.^[1] In contrast, an isolated congenital tricuspid cleft is a rare condition.^[1-3]

The so-called ostium primum defect is an AV septal defect found in a common AV junction, but in which the fused bridging leaflets of the common AV valve are also fused to the scooped-out crest of the ventricular septum. As a result, shunting across the AV septal defect occurs only at atrial level.^[4] These leaflets, irrespective of whether they guard a common AV orifice or separate orifices for the right and left ventricles, bear scant resemblance to the arrangement of the leaflets of the normal mitral and tricuspid valves.^[4]

In the currently accepted definition of the cardiac morphology, it is true that the partial AV canal defect

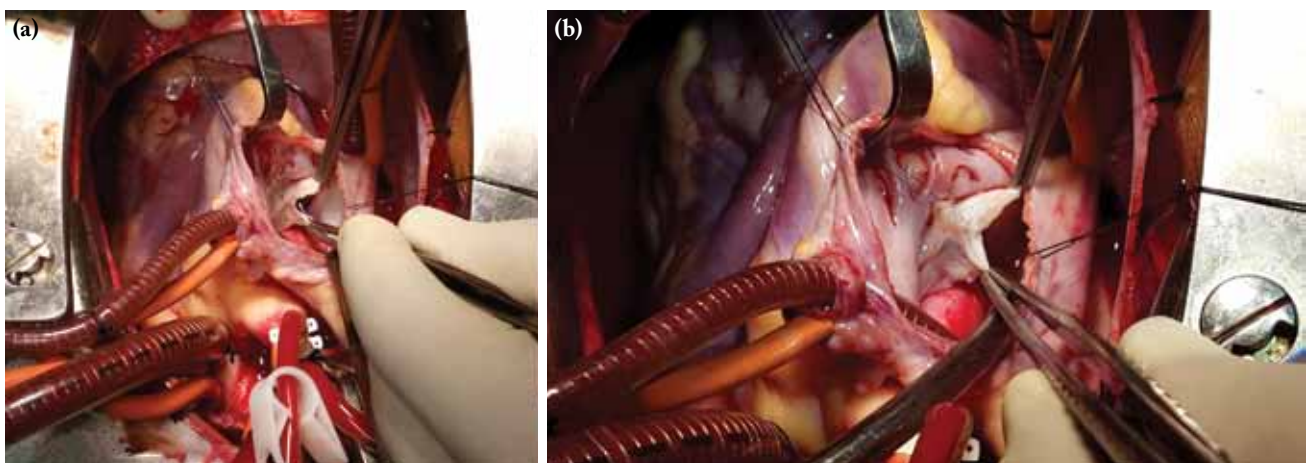


Figure 2. Intraoperative pictures. (a) Complete cleft of a right atrioventricular valve. (b) A suture closed cleft.

will have two separate AV valves although they will not always be well defined. The right AV valve can have three or four cusps depending on the degree of fusion between the anterior and posterior bridging leaflets. If these leaflets are completely fused we can observe a “right atrioventricular valve” made of three cusps namely the fused superior and inferior bridging leaflet, the right anterosuperior leaflet and the right mural leaflet.^[4] Unlike with the cleft left AV valve, the cleft right AV valve does not produce severe symptoms because of the low pressure of the right ventricle and so this condition is often underdiagnosed and it can be associated with other congenital anomalies.^[1-3]

Eichhorn et al.^[1] first reported a 0.6% incidence of right AV valve cleft in patients with a diagnosis of congenital heart disease, and a 0.018% isolated cleft causing right AV valve regurgitation. Embryologically, the right AV valve develops from the inner wall of the right ventricle, by a process called undermining. Eichhorn et al.^[1] described two theories. First, the right AV valve cleft, develops where the lateral endocardial cushion meets the right dorsal conus swelling. Therefore, the cleft might be the result of an anomalous fusion. Second, defective undermining process lead to variable anatomic structure of the right AV valve which may lead to the formation of cleft. Variability makes it difficult to differentiate a cleft from additional valves^[1,2]

The anomalous fusion theory is the most widely accepted theory for complete cleft especially when a partial AV canal defect or endocardial cushion defect is present. The clinical features depend on the severity of tricuspid regurgitation and the associated congenital cardiac anomalies. Most patients commonly have right heart failure and supraventricular arrhythmias. At cardiac auscultation, a grade 2-3/6 holosystolic murmur is audible along with evidence of an associated cardiac malformation. The electrocardiography often demonstrates a right heart volume overload or right bundle branch block, or both. In most cases, the chest X-ray film shows marked enlargement of right-sided heart chambers without signs of pulmonary venous congestion. An echocardiography is the diagnostic tool used for this rare anomaly. Two and three dimensional transthoracic as well as transesophageal echocardiography can delineate this anomaly.^[3]

The most common findings are a visible cleft in the right AV valve and a regurgitation jet in well aligned right AV valve leaflets. If the alignment deformity or prolapse is visible, it may be associated with a congenitally malformed valve and a cleft is very difficult to find in such a scenario. In this case, surgical closure of the cleft with or without a suture or a ring annuloplasty is the treatment of choice.^[1,2] The ultimate decision depends on the severity of the right AV valve regurgitation and the associated congenital anomaly. Isolated mild AV valve regurgitation doesn't require any surgical treatment but the mild regurgitation associated with congenital cardiac anomaly requires correction of the anomaly with the closure of the cleft. For moderate or severe regurgitation some sort of annuloplasty technique will help in the long-term results.

In conclusion, the cleft of the right atrioventricular valve is a rare congenital anomaly. It is probably a result of a malformation of the right atrioventricular valve which occurred during embryologic morphogenesis. The diagnosis is easily made with an echocardiography. In most cases, the treatment of choice is surgical reconstruction of the deficient leaflet with or without suture or ring annuloplasty.

Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding

The authors received no financial support for the research and/or authorship of this article.

REFERENCES

1. Eichhorn P, Ritter M, Suetsch G, von Segesser LK, Turina M, Jenni R. Congenital cleft of the anterior tricuspid leaflet with severe tricuspid regurgitation in adults. *J Am Coll Cardiol* 1992;20:1175-9.
2. Motoyoshi N, Tofukuji M, Sakurai M, Ohmi M, Tabayashi K. Cleft on tricuspid anterior leaflet. *Ann Thorac Surg* 2001;71:1350-1.
3. Séguéla PE, Roubertie F, Tafer N, Thambo JB, Iriart X. Congenital isolated cleft of the tricuspid valve. *Eur Heart J Cardiovasc Imaging* 2014;15:1425.
4. Anderson RH, Baker EJ, Redington A, Rigby ML, Penny D, Wernovsky G, editors. *Pediatric Cardiology*. 3rd ed. Philadelphia: Churchill Livingstone; 2010.