Cardiol Young 2004; 14: 550–552

ISSN 1047-9511

Brief Report

Cleft mitral valve in transposition with intact ventricular septum

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Abstract We describe two cases of an isolated cleft of the mitral valve in transposition with intact ventricular septum. The cleft is positioned leftward in the pulmonary leaflet, at about 2 o'clock, when viewed from below looking at the cardiac short axis with the right ventricle to one's left hand. Such clefts, when seen in the Taussig-Bing malformation are also positioned leftward.

In keeping with our current knowledge of cardiac development, our cases provide further evidence that transposition with an intact ventricular septum is the end-point of the Taussig-Bing spectrum.

Keywords: Discordant ventriculo-arterial connections; Taussig-Bing malformation; trifoliate left atrioventricular valve

THE SO-CALLED "CLEFT" NOTED IN THE LEFT atrioventricular valve of patients with an atrioventricular septal defect and common atrioventricular junction is a misnomer. In reality, it is the zone of apposition between the superior and inferior bridging leaflets of the common atrioventricular valve.[1,2] When viewed from below, looking up the cardiac short axis with the right ventricle to one's left hand, the so-called "cleft" is observed medially between 9 and 10 o'clock. When a true cleft of the anterior or aortic leaflet of the normal mitral valve is viewed in similar fashion, it occupies a position at 11 o'clock.[3–5] (Fig. 1). In the Taussig-Bing malformation, however, should the forming mitral valve retain a cleft between its component parts, the cleft will be positioned more leftward.[6,7] The essence of the Taussig-Bing malformation, of course, is the presence of a subpulmonary interventricular communication. We have now encountered two children who had an arterial switch in the newborn period for transposition with an intact ventricular septum. Both were observed to have an isolated cleft in the pulmonary leaflet at the mitral valve, but surprisingly, the cleft was positioned even more leftward, at about 2 o'clock.

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Accepted for publication 18 May 2004

Case reports

LB presented cyanosed on the first day of life, and was found to have transposed arterial trunks with an intact ventricular septum. Following an atrial balloon septostomy, he came forward to an uneventful arterial switch on the seventh day. He developed a supraventricular tachycardia with aberrancy on the 19th day, but responded well to intravenous adenosine, and remained symptom-free on twice-daily Sotalol, eventually being weaned from that medication 8 months later. His subsequent course was complicated by increasing incompetence of his neoaortic valve, associated with mild-to-moderate incompetence of the mitral valve. The latter was eventually shown to be due to an isolated cleft, positioned at about 2 o'clock when viewed from below looking at the cardiac short axis with the right ventricle to the left (Fig. 2). By the age of 4 years, he developed increasing left ventricular dilation initially helped by oral Lisinopril. His echocardiogram showed left ventricular hypertrophy. With continuing decrease in exercise tolerance and a dilated left ventricle with left

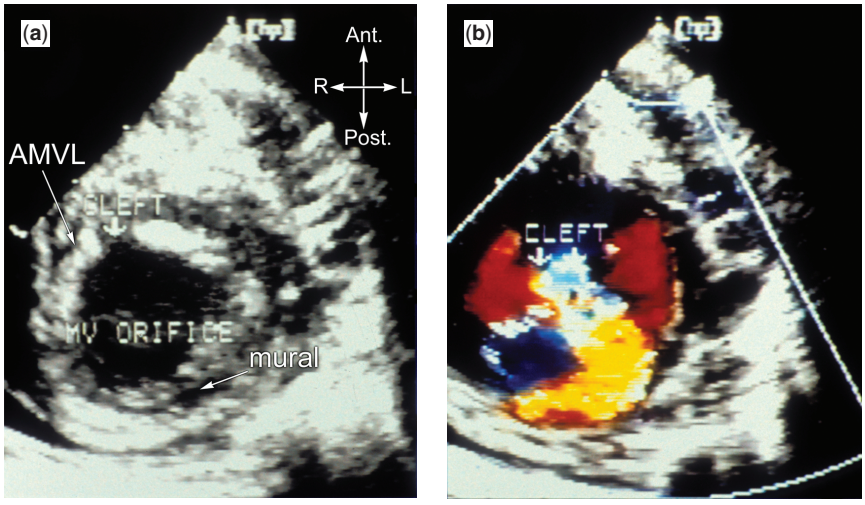

Figure 1.
Parasternal short axis of left ventricle of a normal heart showing an isolated cleft in the anterior mitral valve leaflet positioned at 11 o'clock. The isolated cleft divides the aortic leaflet of the mitral valve into 2 segments in diastole. Colour Doppler shows the regurgitant jet through the cleft.

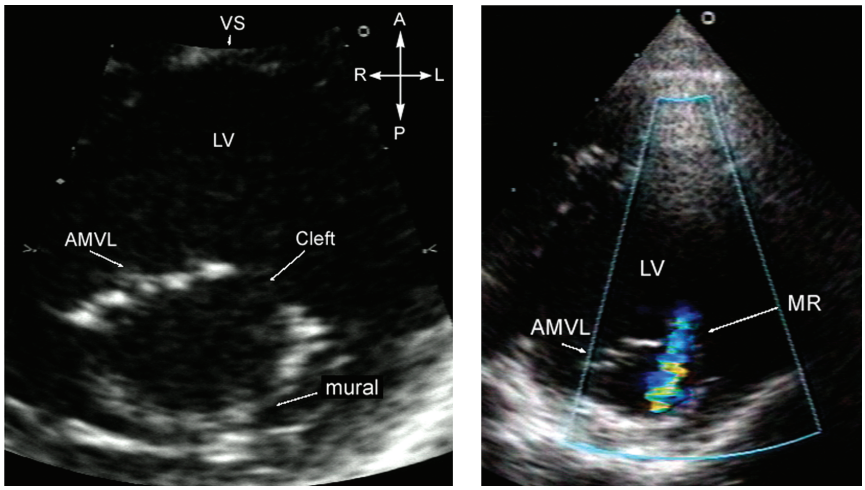

Figure 2.
Parasternal short axis of the left ventricle showing a cleft in the pulmonary leaflet of the mitral valve positioned at 2 o'clock in the setting of transposition and intact ventricular septum. VS: ventricular septum; LV: left ventricle; AMVL: anterior mitral valve leaflet; MURAL: mural leaflet of mitral valve; MR: mitral regurgitation; MV: mitral valve. [*]*Reprinted from: Tamura M, Menahem S, Brizard C. Clinical features and management of cleft mitral valve in childhood. J Am Coll Cardiol 35: 764–770. Copyright 2000 with permission from the American College of Cardiology Foundation.*

ventricular function at the lower limit of normal, he now has been accepted at the age of 6 years for repair of his neoaortic valve and suture of the cleft mitral valve.

DF presented on the first day of life with cyanosis, initially attributed to meconium aspiration. Cross-sectional echocardiography revealed transposed arterial trunks with an intact ventricular septum. A balloon atrial septostomy was followed 5 days later by an uneventful arterial switch. He has remained well subsequently. His neopulmonary valve, however, was noted to be bicuspid, initially producing mild-to-moderate stenosis, for which he required no treatment. Subsequent echocardiograms revealed increasing mitral incompetence. Careful review of the valve showed an isolated cleft in a similar position as the previous patient. At the age of 11 years, he came forward to balloon valvoplasty of his moderately stenosed neopulmonary valve, with a drop of his right ventricular pressure from just over two-thirds systemic pressures to just under half systemic pressures. He currently remains well at the age of 14, with evidence of mild pulmonary valvar stenosis and mild mitral incompetence. There is no immediate plan to attend to his cleft mitral valve.

https://doi.org/10.1017/S1047951104005128 Published online by Cambridge University Press

Discussion

As far as we are aware, isolated clefts of the mitral valve have not been reported in transposition with intact ventricular septum. Our observations, nonetheless, are in keeping with our current knowledge of cardiac development. Thus, the Taussig-Bing lesion represents a spectrum between double outlet right ventricle with subpulmonary defect and discordant ventriculo-arterial connections with a malalignment ventricular septal defect. This spectrum shows us that, when the heart has discordant ventriculo-arterial connections, the pulmonary trunk enters the right ventricle through the left side of the ventricular outflow tract, with the outlet septum fusing with the muscular ventricular septum to wall the pulmonary trunk, rather than the aorta, into the left ventricle.[8] In the process of normal development, the outlet septum fuses antero-superiorly relative to the interventricular communication, thus walling the aorta into the left ventricle. This arrangement gives us the spectrum seen with double outlet and subaortic defect, of which tetralogy of Fallot is part.

In the setting of discordant ventriculo-arterial connections with intact septum, therefore, the pulmonary outflow tract is positioned much more leftward in comparison to the subaortic outflow tract of the normal heart, and is not "wedged" between the mitral valve and the septum to the same extent as is the normal subaortic outflow tract.

Thus, when the forming mitral valve retains a cleft between its component parts, this cleft will also be positioned more leftward, as is seen with straddling of the mitral valve, also often cleft, in the Taussig-Bing malformation.[6,7]

The location of the cleft in our two cases, therefore, is in keeping with our current knowledge of cardiac development. It provides additional evidence that transposition with an intact ventricular septum is, indeed, the end-point of the Taussig-Bing spectrum.

In both of our cases, surprisingly, retrospect review of the earlier echocardiograms failed clearly to demonstrate the cleft. Had there been an awareness of the possible association, and a more careful study undertaken at the time, it may have led to an earlier diagnosis and possible correction at the time of the initial surgery.

References

1. Smallhorn JF, Tommasini G, Anderson RH, Macartney FJ. Assessment of atrioventricular septal defects by 2 dimensional echocardiography. Br Heart J 1982; 47: 100–121.
2. Sigfusson G, Ettedgni JA, Silverman NH, Anderson RH. Is a cleft in the anterior leaflet of an otherwise normal mitral valve an atrioventricular canal malformation? J Am Coll Cardiol 1995; 26: 508–515.
3. Di Segni E, Bass JL, Lucas RV, Einzig S. Isolated cleft mitral valve: a variety of congenital mitral regurgitation identified by two dimensional echocardiography. Am J Cardiol 1983; 51: 927–931.
4. Smallhorn JF, de Leval M, Stark J, Sommerville J, Taylor JF, Anderson RH, Macartney FJ. Isolated mitral cleft: two dimensional echocardiographic assessment and differentiation from "clefts" associated with atrioventricular septal defects. Br Heart J 1982; 48: 108–116.
5. Tamura M, Menahem S, Brizard C. Clinical features and management of isolated cleft mitral valve in childhood. J Am Coll Cardiol 2000; 35: 764–770.
6. Soto B, Ceballos R, Nath PH, Bine RM, Pacifico AD, Begeron LM. Overriding AV valves, an angiographic–anatomic correlate. Int J Cardiol 1985; 9: 327–339.
7. Stellin G, Zuberbuhler JR, Anderson RH, Siewers RD. The surgical anatomy of the Taussig-Bing malformation. J Thorac Cardiovasc Surg 1987; 93: 560–569.
8. Smith A, Wilkinson JL, Anderson RH, Arnold R, Dickinson DF. Architecture of the ventricular mass and atrioventricular valves in complete transposition with intact ventricular septum compared with the normal 1: the Left ventricle, mitral valve and interventricular septum. Pediatr Cardiol 1986; 6: 253–257.