

The Abnormal Contralateral Atrioventricular Valve in Mitral and Tricuspid Atresia in Neonates: An Echocardiographic Study

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Abstract. Abnormalities of the mitral valve (MV) or the tricuspid valve (TV) morphology and/or function in patients with functional single ventricle may result in early morbidity and death. The purpose of this study was to determine the incidence of contralateral atrioventricular valve (AVV) pathologies in mitral valve atresia (MA) and tricuspid valve atresia (TA). We retrospectively reviewed the echocardiographic data of 50 neonates with MV and 20 with TA. Appearance of the papillary muscles, chordae tendinae, and valve leaflets was assessed. AVV regurgitation was semiquantitated by color-flow Doppler and the AVV annulus diameter was measured and indexed to body surface area. MV abnormalities were found in 9 of 20 (45%) of patients with TA. The MV was myxomatous in 9 patients, the leaflets were redundant in 5 patients, and prolapsing occurred in 4 patients. Mild regurgitation was found in 2 patients. In 18 of 20 (90%) patients MV annulus size was larger than 95% of predicted normal values. TV abnormalities were found in 12 of 50 (24%) patients with MA. The TV was myxomatous in 4 patients, prolapsing in 2, and redundant in 3, and moderate TV regurgitation was found in 3 patients. In 29 of 50 (58%) patients TV annulus size was larger than 95% of predicted normal values. Contralateral AVV abnormalities in tricuspid and mitral valve atresia are common and should be assessed carefully before surgical procedures.

Key words: Functional single ventricle — Fontan procedure — Contralateral atrioventricular valve pathologies

One of the most important parameter to be assessed in patients with univentricular hearts being evaluated for a possible Fontan operation is the status of the atrioventricular valves [2, 5]. Earlier studies of the normal tri-

cuspid and mitral valve showed numerous variations of valve morphology [13, 14]. In Fontan patients gross pathology can be a disqualifying cause for surgery. Abnormalities of the mitral or tricuspid valve morphology and/or function may result in early morbidity and death. Sporadic cases of pathology regarding the mitral valve in patients with tricuspid atresia have been reported [10, 15]. Bharati and Lev [3] stated in 1979 that the mitral valve in the tricuspid atresia complex most often shows little abnormality. In contrast to previous reports, a study on an autopsy material was published in 1989 where a high incidence of valvar pathology was described. In 31 autopsy specimens with right-sided tricuspid atresia, 16 cases (52%) with unequivocal pathology of the mitral valve were noted [9].

Since then the number of patients undergoing surgery for mitral atresia (as a subgroup of hypoplastic left heart syndrome) has increased markedly. The status of the tricuspid valve in these patients is as important as that of the mitral valve in tricuspid atresia. We are not aware of any echocardiographic study analyzing the incidence of valvar pathology in these two groups of patients. The purpose of this study was to determine the incidence of different types of contralateral atrioventricular valve pathologies in mitral and tricuspid valve atresia.

Patients

A total of 70 consecutive neonates with tricuspid valve atresia or mitral valve atresia were included in the study. In 50 cases the diagnosis was mitral atresia, and 20 patients were diagnosed as tricuspid atresia. There were 38 male and 32 female neonates with a mean age of 4.8 days (range 1–24 days).

Methods

Two independent observers reviewed echocardiographic and Doppler data from routine studies. Disagreements were solved by consensus. The following parameters were measured and evaluated:

1. Atrioventricular valve regurgitation was semiquantitated by color-flow Doppler imaging, taking into consideration length of jet intrusion into the receiving atrium, proximal jet width, proximity of the jet to adjacent walls, and driving pressure [4].
2. Atrioventricular valve diameter was measured in short-axis, apical, and subcostal four-chamber view. The projection with the largest diameter was chosen and the mean of three beats was used. The diameter was indexed to body weight and compared to established normal values [12].
3. Leaflet morphology
4. Appearance of the papillary muscles and the length of the chordae

Results

Tricuspid Atresia with Mitral Valve Abnormalities

Mitral valve abnormalities were found in 9 of 20 (45%) patients with tricuspid atresia. The indexed mitral valve annulus diameter was larger than 95% of the predicted value in 18 patients (90%). Mild mitral valve regurgitation was noted in 2 patients (10%) and myxomatous mitral valve was found in 9 patients (45%). Redundant valve was seen in 5 patients (25%) and mitral valve prolapse in 4 patients (20%). Four patients (20%) had an isolated cleft, and malattachment of the chordae was noted in 3 patients (15%).

Mitral Atresia with Tricuspid Valve Abnormalities

Tricuspid valve abnormalities were found in 12 of 50 (24%) patients with mitral atresia. The indexed tricuspid valve diameter was larger than 95% of predicted value in 29 patients (58%). Mild tricuspid valve regurgitation was found in 16 of 50 patients (32%) and moderate tricuspid valve regurgitation was present in 3 patients (6%). The tricuspid valve was myxomatous and redundant in 4 (8%) and 3 (6%) patients, respectively. Tricuspid valve prolapse was found in 2 patients, and in 4 patients malattachment of the chordae was seen. All results are summarized in Table 1.

Discussion

In patients with tricuspid atresia and mitral atresia, the circulation is dependent on one pumping ventricle with one atrioventricular valve. One of the most important parameter to be assessed in patients with univentricular hearts being evaluated for a possible Fontan operation is the status of the atrioventricular valve [5]. The function and the morphology of the single atrioventricular valve in tricuspid and mitral atresia is essential for a successful final repair for these malformations. Earlier studies have shown that patients undergoing Fontan repair with atrioventricular valve insufficiency requiring repair or replacement are at higher risk [7]. This is also true for patients with hypoplastic left heart syndrome [2].

Table 1. Pathologic findings in the contralateral valve in tricuspid atresia and mitral atresia

Parameter	Tricuspid atresia	Mitral atresia
	Mitral valve, <i>n</i> = 20	Tricuspid valve, <i>n</i> = 50
Indexed valve diameter >95% of predicted value	90%	58%
Myxomatous valve	45%	8%
Redundant leaflets	25%	6%
Valve prolaps	20%	4%
Chordal malattachment	15%	8%
Isolated cleft	20%	
Valve regurgitation	10% (mild)	32% (mild) 6% (moderate)

This study shows that contralateral atrioventricular valve abnormalities in tricuspid and mitral valve atresia are common. The findings regarding tricuspid atresia and the mitral valve in this echocardiographic study are similar to those described by Ottenkamp and Wenink [9] in their study based on analysis of autopsy specimens with right-sided tricuspid atresia. We found the same spectrum of pathology and a similar incidence. Our study shows that the incidence of tricuspid valve pathology in mitral atresia is considerably lower than that in the mitral valve pathology in tricuspid atresia.

Besides the strict morphogenetic theories [1, 16], a possible hemodynamic explanation for the more frequent finding of an enlarged mitral valve in tricuspid atresia, compared to the tricuspid valve in mitral atresia, can be applied. In the normal fetus, the left ventricle handles smaller volumes than the right ventricle. Studies on fetal lambs have shown that the right ventricle receives and ejects two-thirds of the combined ventricular output [6]. Recent Doppler flow evaluation studies have also suggested a hemodynamic dominant right ventricle, although its dominance is less than that described [8]. Significant hemodynamic disturbances occur with tricuspid and mitral anomalies, diverting blood flow into the contralateral ventricular chamber [11]. These chambers enlarge, consequently enlarging the atrioventricular annulus. The tricuspid valve could be predisposed to handle a significantly larger flow volume than the mitral valve, and could be less vulnerable to annulus enlargement.

Conclusion

Contralateral atrioventricular valve abnormalities in tricuspid and mitral valve atresia are common, most common in tricuspid atresia, and should be assessed carefully before surgical procedures.

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