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Absent Conal Septum in Tetralogy of Fallot

An Angiographic Study

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ABSTRACT

Absent conal septum in tetralogy of Fallot (TF) is usually noted intraoperatively when the ventricular septal defect (VSD) is found abutting the pulmonary valve, its superior rim being nearly inaccessible transatrially, and the posterior rim being separated from the tricuspid valve (TV) by a muscular ridge. The authors retrospectively analyzed angiograms of 208 consecutive patients with TF seen at their center from July 1989 to December 1995 for absence of the conal septum and the presence of an interval between the TV and the margin of the VSD in 30° right anterior oblique view. In 13 (6%) patients, angiograms were inadequate or of poor quality for assessment and were excluded. Twenty-two of the remaining 195 (11%) patients had a large muscular interval between the tricuspid annulus and the margin of the VSD, which was associated with an absent conal septum in 14 (7.2%) and a diminutive septum in 8 (4%) patients. Nine of the 14 patients with an absent conal septum at angiography underwent surgery, and this finding was confirmed in all. The authors conclude that absent conal septum is not uncommon in TF and constitutes an important variation in its anatomy that can be identified preoperatively at angiography for optimal surgical management.

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Introduction

Tetralogy of Fallot (TF) is characterized by anterior and leftward deviation of the conal (infundibular) septum,[1] a large nonrestrictive malalignment ventricular septal defect (VSD), right ventricular (RV) hypertrophy, and varying degrees of pulmonary arterial hypoplasia.[2] The VSD is classically perimembranous (subaortic) in position, bounded posteroinferiorly by the tricuspid annulus and anterosuperiorly by the hypertrophied conal (infundibular) septum.[2-4] In a minority, the conal septum is absent, the associated VSD being both subaortic and subpulmonary (subarterial) with its inferior margin formed by a remnant of the membranous septum (total conus defect) or a muscular ridge (subtotal conus defect) and its superior margin extending to abut against the aortic and pulmonary annuli.[5]

This morphologic entity has usually been detected intraoperatively and has been reported most frequently from South America[6,7] and the Far East[5,8-11] with only a few cases reported from other parts of the world.[12,13] Unrecognized, it constitutes an important cause of mortality after intracardiac repair,[5-9] and its detection preoperatively has been emphasized to facilitate optimal surgical management.[5,8]

Right ventricular angiography in the straight right anterior oblique (RAO 30°)[14] or elongated right anterior oblique (RAO 30° with cranial 30° tilt)[10,12] has been suggested to optimally define the anatomy of the RV infundibular segment and demonstrate the absence of the conal septum.

We aimed to determine the frequency of absent conal septum in Indian patients with TF and study whether this entity could be detected preoperatively by using angiographic analysis.

Patients and Methods

We studied, retrospectively, 208 consecutive patients with TF undergoing cardiac catheterization and angiography at our center from July 1989 to December 1995.

The right ventricular (RV) angiograms of all patients were analyzed. Angiograms were included only if a straight RAO projection of the RV angiogram was available. Angiograms of poor quality, unsuitable for interpretation, were excluded. All angiograms were reviewed independent of surgical data.

The RV angiograms were analyzed for the presence of (1) a muscular ridge, seen as an interval, between the tricuspid valve annulus and the VSD; and (2) the conal septum, seen as a negative shadow, separating the pulmonary and aortic outflow tracts. Based on the angiographic findings, the VSD in each case was classified into three types:[12] (1) perimembranous, ie, no muscular interval between the TV and the VSD with a hypertrophied conal septum identifiable (Figure 1); (2) muscular outlet, ie, presence of a muscular interval between the TV and the VSD, and an identifiable, though diminutive, conal septum (Figure 2); and (3) Subarterial, ie, presence of a large muscular interval between the TV and the VSD, but no conal septum identifiable (Figure 3).

The absence of the conal septum was noted and the type of VSD was identified during intracardiac repair. The surgical findings were correlated in a blinded fashion with preoperative angiographic findings by independent observers.

Results

Angiographic Results

A total of 208 patients with TF had undergone preoperative cardiac catheterization and angiography during the study period. Thirteen (6%) angiograms were excluded from analysis (3 owing to poor quality, 6 because of unavailability of an RAO view, and 4 because they could not be retrieved).

Of the remaining 195 patients whose angiograms were reviewed, 22 (11%) had a large muscular interval between the TV and the VSD. This was associated with absence of the conal septum in 14 (7.2%) and a diminutive conal septum in 8 (4%). In all patients with an absent conal septum, the pulmonary stenosis was primarily annular, although 8 of them had varying degrees of valvular pulmonary stenosis in addition.

Surgical Results

Of the 208 patients catheterized, 101 (48%) underwent intracardiac repair at our center by December 1995. Among these surgical patients, 9 were identified as having absence of the conal septum at surgery. This could be confirmed in the preoperative angiographic analysis in all. No distinction could be made at surgery between pa-

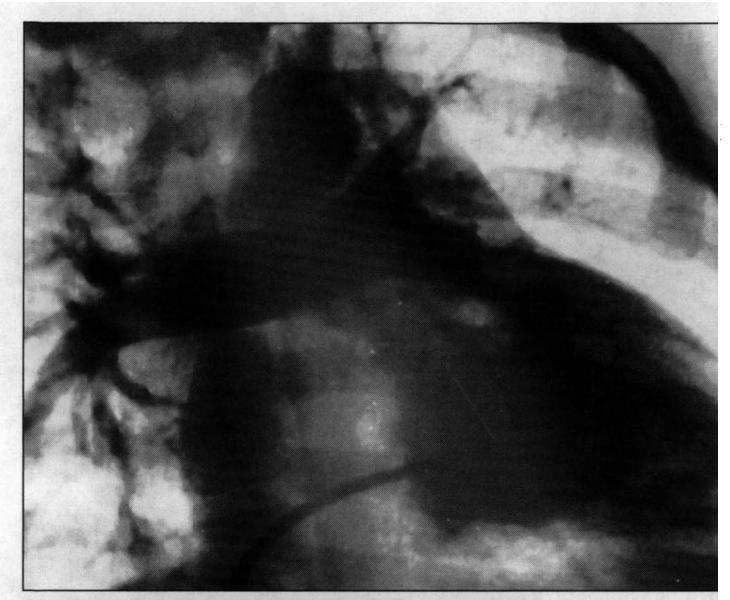

Figure 1. Straight right anterior oblique (RAO 30°) projection of the right ventricular angiogram showing absence of a muscular interval between the tricuspid valve and the ventricular septal defect, with a hypertrophied conal septum.

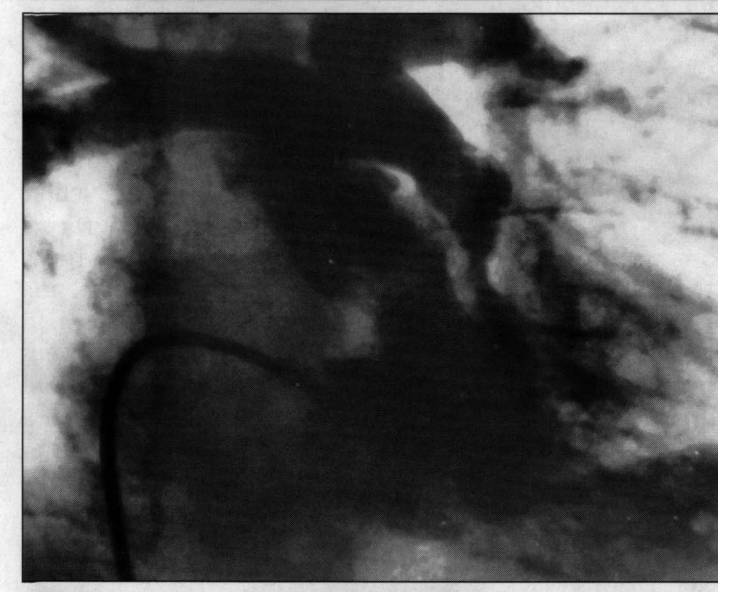

Figure 2. Straight right anterior oblique (RAO 30°) projection of the right ventricular angiogram showing a muscular interval between the tricuspid valve and the ventricular septal defect and a diminutive conal septum.

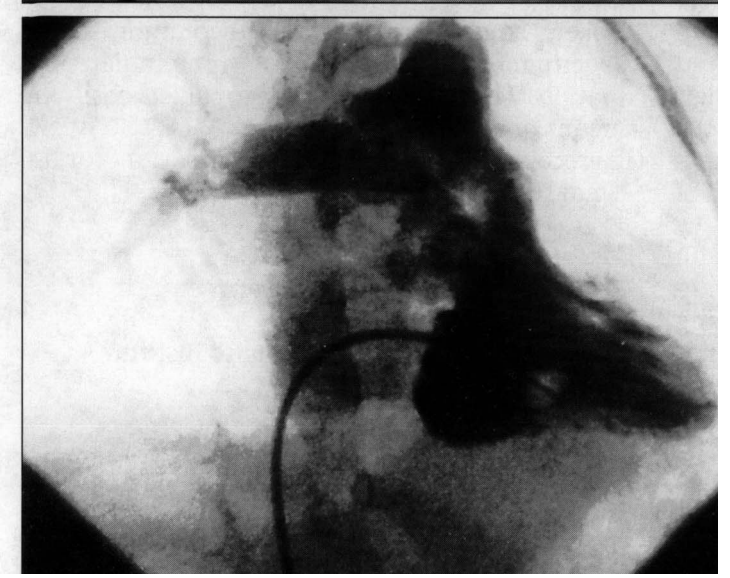

Figure 3. Straight right anterior oblique (RAO 30°) projection of the right ventricular angiogram showing a large muscular interval between the tricuspid valve and the ventricular septal defect and an absent conal septum.

tients with a diminutive conal septum and those with classical TF. In addition, no patient was found to have an absent conal septum at surgery when the same was not identified on the preoperative angiogram. All patients with an absent conal septum needed a combined transatrial transpulmonary approach for VSD closure. A right ventricular outflow patch was required in 8 of the 9 (89%) patients with this anatomy.

Discussion

Absent conal septum has been recognized to constitute an important variation in the morphologic spectrum of TF in South America[6,7] and the Far East.[5,8-11] In our study of Indian patients, this entity constituted 7.2% of all cases studied angiographically and 9% of all patients operated on. Similar figures were reported by Ando from Japan (9.3%)[5] and Vargas et al from Argentina (6.6%)[7] among patients with TF who were operated on.

Angiographic Analysis

Preoperative analysis of RV angiograms in the straight RAO (RAO 30°) view optimally demonstrated the absence of the conal septum and the presence of a muscular interval between the tricuspid valve and the margin of the VSD, characteristic of this entity. We could confirm our angiographic findings in all such patients who underwent intracardiac repair.

Although straight RAO (RAO 30°)[14] and elongated RAO (RAO 30° with 30° cranial tilt)[10,12] projections have both been suggested in the diagnosis of this entity, Soto et al[12] described only 1 patient with an absent conal septum, and Chen et al,[10] using an elongated RAO view, could achieve a predictive accuracy of only 73% in their study of 30 patients with tetralogy of Fallot. We feel, therefore, that a straight RAO view is superior to an elongated RAO view in the diagnosis of this entity as demonstrated by our high accuracy of angiographic detection, and this is based on the premise that a cranial tilt given to an RV angiogram in an RAO projection starts visualizing the outflow septum enface[15] and, hence, may not profile it well.

Surgical Implications

Identification of an absent conal septum is important preoperatively since the anatomy of the infundibular region in this entity differs markedly from that in classical TF.[5,7,12] The VSD is positioned "high," and the aorta is more severely dextroposed. This results in the superior rim of the VSD being almost invisible and inaccessible transatrially[16] except with extreme traction of the TV[17] with the attendant risk of damage to the tricuspid valve. This necessitates a combined transatrial and transpulmonary approach, or a transventricular approach for repair.[16,17] All our patients required a combined approach for VSD closure.

Absence of the conal septum necessitates placing sutures of the VSD patch through the thin fibrous rim at the zone of pulmonic and aortic valvar continuity, with risk of damage to the aortic valve and subsequent surgically induced regurgitation.[7]

In earlier studies, a high mortality rate of up to 50% was reported after intracardiac repair of this entity.[5,6,8,9] Vargas et al[7] observed that the VSD patch combined with the hypertrophied RV walls and the markedly dextroposed aorta invariably produced narrowing of the right ventricular outflow tract, resulting in a high postoperative mortality rate. Using a right ventricular outflow tract (RVOT) patch in almost all their patients, they were able to reduce the mortality rate to 4.3%. In our study, 8 of the 9 patients with absent conal septum needed an RVOT patch for adequate relief of pulmonary stenosis. The need for such a patch may also mean that neonatal repair is not the appropriate surgical strategy for this entity.[16]

Conclusion

Absent conal septum is not uncommon in the spectrum of tetralogy of Fallot in the Indian population. This entity can be recognized easily by means of an RV angiogram in the RAO 30° view. Its recognition preoperatively is important for optimal surgical management.

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