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Surgical Repair of Infundibular Ventricular Septal Defect and Aortic Regurgitation

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Interpretation of the anatomy of the defect and the pathophysiology has guided the surgical technique and indications for infundibular ventricular septal defect VSD. Infundibular ventricular septal defects are located in the infundibular septum, between the two commissures of the right coronary cusp. The defect is associated with an anomaly of the right sinus of Valsalva where the transition to cusp tissue occurs higher than normally. There is development of fibrous adherences between the ventricular surface of the right coronary cusp and the crest of the septum. This reduces the height of the cusp and destabilizes the valve. The aim is to reposition the hinge point of the right coronary cusp to restore the normal height of the cusp, hence a normal surface of apposition. This is achieved with a trans-aortic approach. The major difference with other techniques described is the extensive mobilization of the cusp. The procedure is completed by a reduction of the free edge of the right coronary cusp if it is elongated. This technique is indicated in all patients with infundibular ventricular septal defect in whom an aortic regurgitation appears or increases during follow-up. Fifteen patients were operated on with this technique between 1996 and 2005. Thirteen have achieved good results at follow-up. There was one late death.
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The technique described herein was devised to treat the specific anatomic condition of the infundibular ventricular septal defect (VSD) with aortic regurgitation. At Royal Children's Hospital, Melbourne, Australia, the anatomy of the defect and our understanding of the pathophysiology, together with the work from previous authors,[1-3] have guided the surgical technique and indications. The defect is significantly more common in Asia, and we have also gained confidence in its approach and understanding of the pathophysiology in the Ho Chi Minh City Heart Institute in Viet Nam.

Anatomy

The technique described in this article was devised to treat what we understand is a specific anatomic condition. Several anatomic types of VSD with connection to the aortic valve are classically described: peri-membranous and conal VSD. Multiple names for conal VSDs are in use. They can also be labeled infundibular VSD, subpulmonary VSD, doubly committed VSD, and subarterial VSD. These multiple denominations are not attached to specifically different anatomic subtypes. We believe that within the conal septum (where the conus is the subpulmonary infundibulum), there are different subtypes of VSD and that these should be named differently.

On the anatomic point of view, within the conal septum, one can encounter (1) an absence of conal septum, usually within a subset of tetralogy of Fallot, where the aortic and pulmonary valve annuli belong to the same plan and each valve provides the support for the other; (2) a high muscular VSD with a muscular rim all around; (3) a circular defect underneath part of the right coronary cusp; and (4) a crescendic defect underneath the right coronary cusp of the aortic valve.

We believe numbers 3 and 4 are separate entities in their natural history and anatomically and deserve different surgical approaches.

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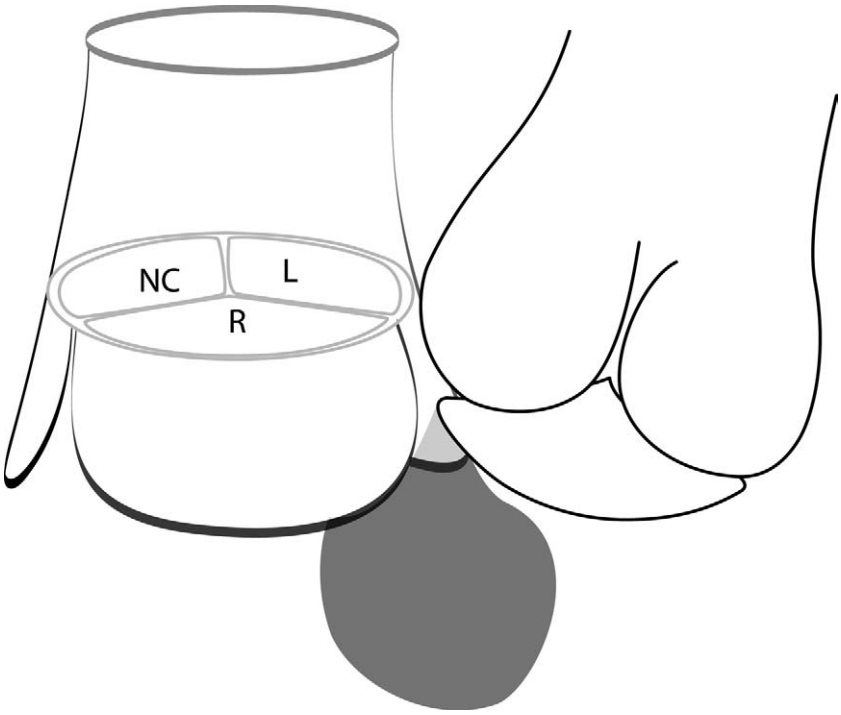

Figure 1 Schematic representation of a large defect in the conal septum with little relationship with the right coronary cusp.

The first is a large circular defect in the conal septum with a short segment of its circumference common to right coronary cusp hinge point, close to the left-right commissure. On the pulmonary valve, the defect has a common border with the left and posterior pulmonary cusps. They present early in the neonatal period with large shunt generating cardiac failure and failure to thrive. The surgical indication is to suppress the shunt. The occurrence of aortic regurgitation is infrequent,[4] as in a perimembranous VSD. The approach is through the pulmonary orifice and the VSD is closed with a patch. We have represented such a VSD in Figure 1.

The second is the one we intend to focus on in this article. At the Royal Children's Hospital, Melbourne we have labeled this defect "infundibular VSD," and we shall do so in the remaining part of this article. It is located in the infundibular septum and is strictly limited between the two commissures of the right coronary cusp. The defect is a lack of fusion of the right sinus of Valsalva to the crest of the septum at the level of the hinge line of the right coronary cusp. This is suggested by the crescent shape of the defect. The normal crescentic shape of the hinge point of the right coronary cusp molds the crest of the septum. The defect seems to be associated with an anomaly of the right sinus of Valsalva where the transition from sinus tissue to cusp tissue is higher than normal, at least when the lesions are seen at the time of the surgical procedure. On the right side of the septum, the pulmonary and aortic annuli belong to separate plans and meet at an angle as in a normal heart. Very little of either circumference is com-

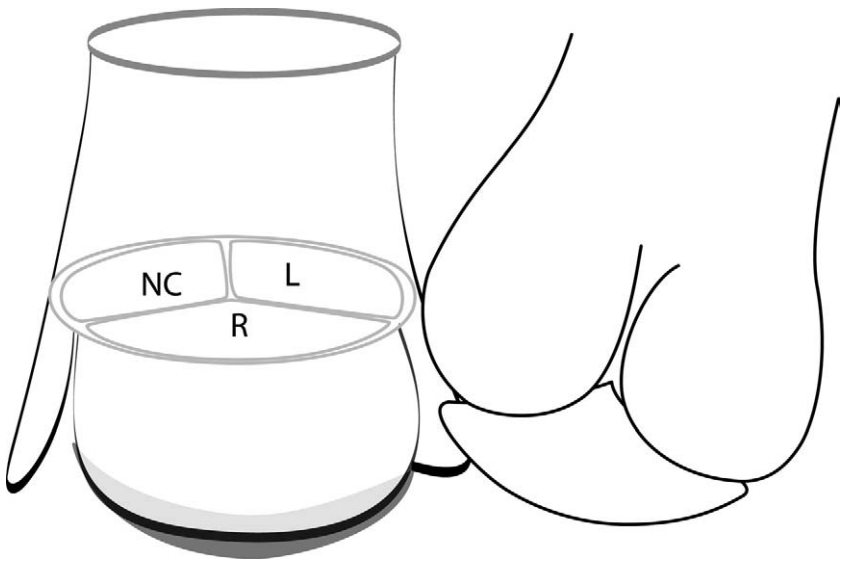

Figure 2 Schematic representation of an "infundibular VSD" as we see it.

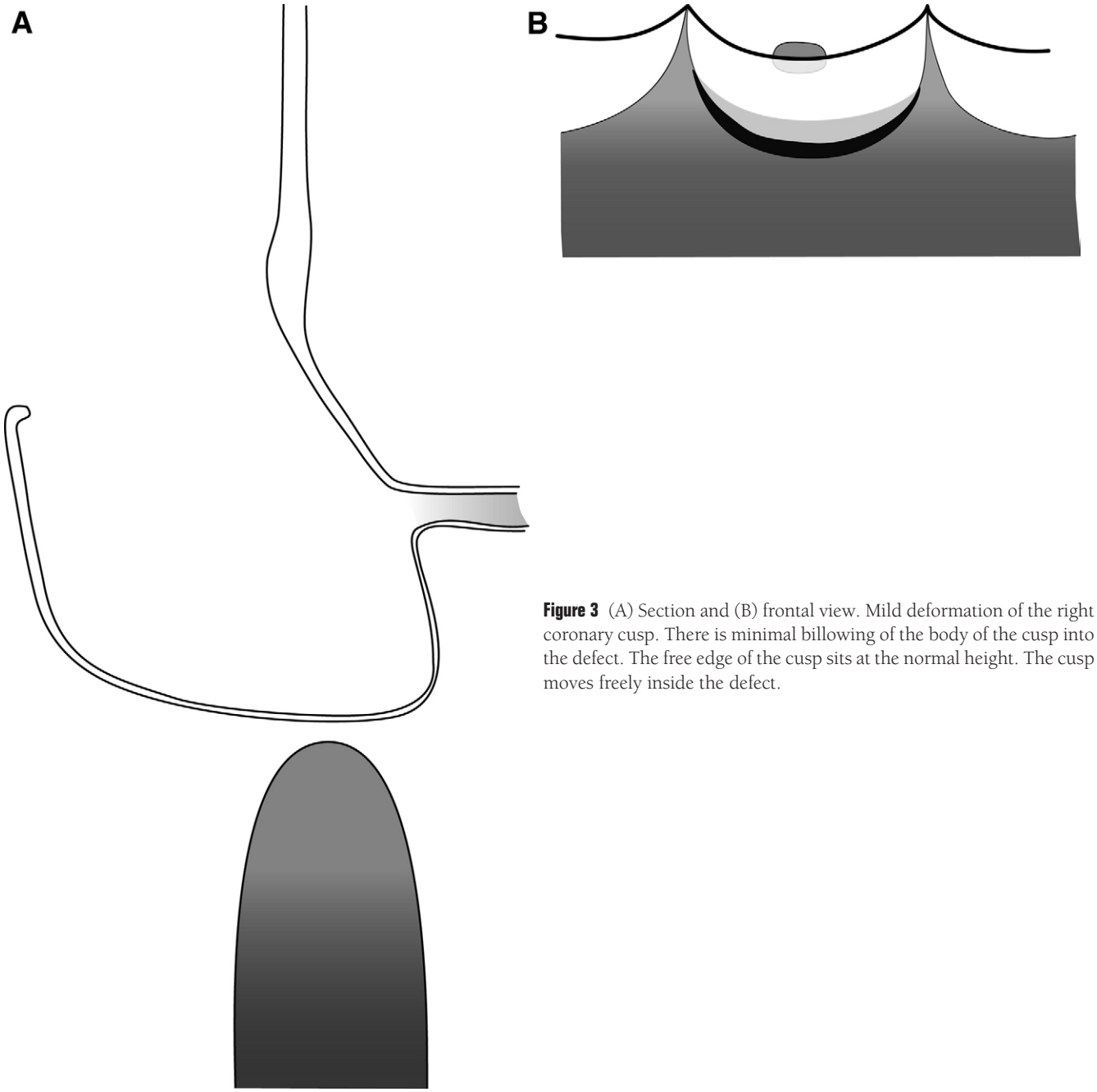

Figure 3 (A) Section and (B) frontal view. Mild deformation of the right coronary cusp. There is minimal billowing of the body of the cusp into the defect. The free edge of the cusp sits at the normal height. The cusp moves freely inside the defect.

mon to both annuli, and virtually no part of the defect is common to the pulmonary cusps hinge points. The defect never generates large shunt. The aortic regurgitation appears progressively. The deformation of the right coronary cusp continues to progress during the life of the patient and is almost constant above 30 years of age[5]; eventually, some degree of aortic regurgitation is constant without surgical treatment. We have represented an infundibular VSD in Figure 2.

Pathophysiology

We believe that pathophysiology also provides grounds for the separation of the two entities. We believe that the Ventury effect[6] plays a limited role in the limitation of the shunt in the infundibular VSD. The infundibular VSD is restrictive before the right coronary cusp has been deformed and prolapses into the defect. This suggests that the anatomy of the infundibular VSD itself is sufficient to limit the shunt and the limitation of the shunt is not a consequence of the secondary deformation of the cusp.

Our interpretation of the pathophysiology for the development of aortic regurgitation in infundibular VSD is the following. At any stage of the presentation, the shunt is small, as the hinge point of the right cusp is covering the area of the defect. We see the mechanism of the creation of the aortic regurgitation as a succession of events.

1. The lack of attachment of the sinus of Valsalva to the septum allows the hinge point of the right coronary

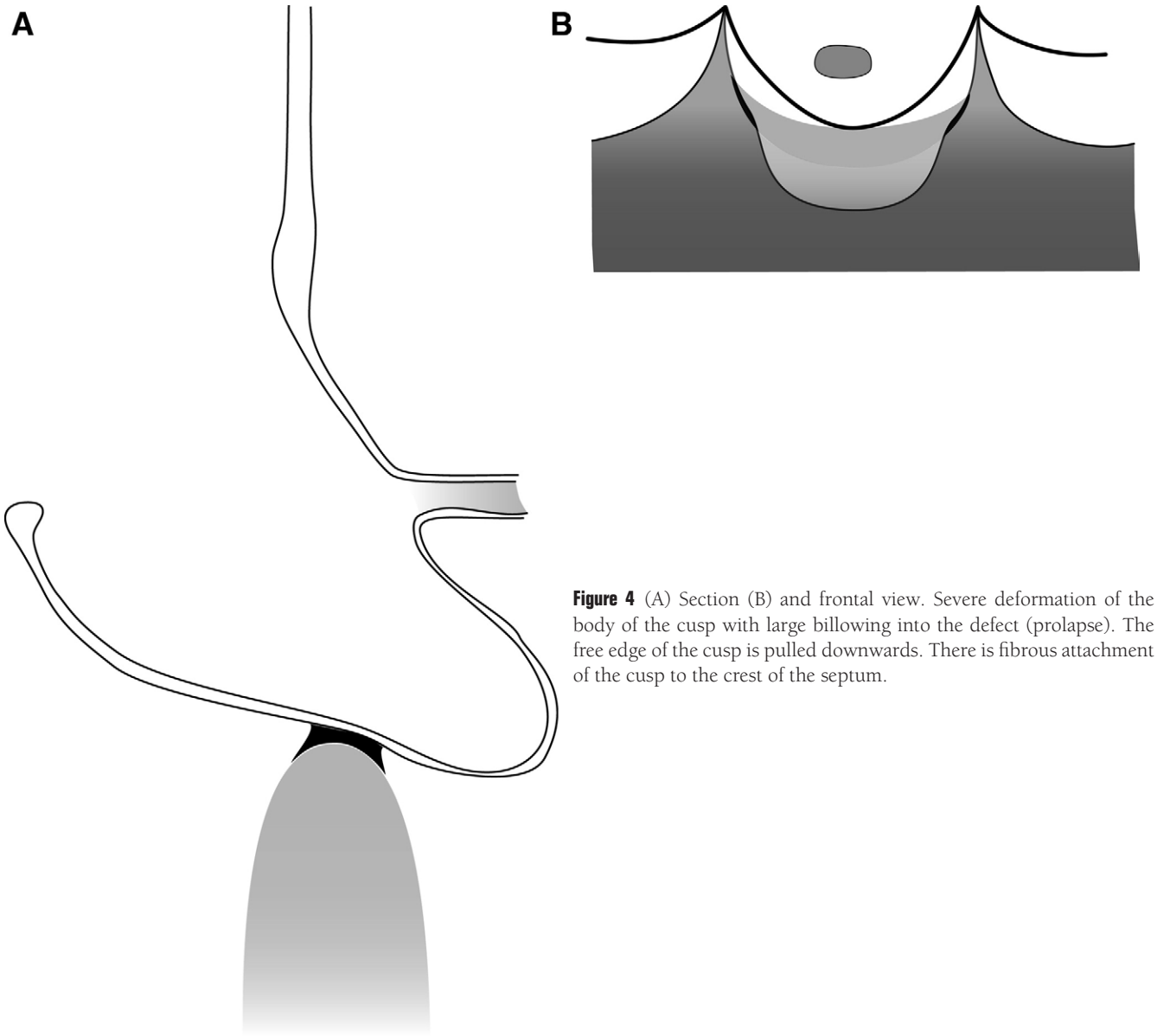

Figure 4 (A) Section (B) and frontal view. Severe deformation of the body of the cusp with large billowing into the defect (prolapse). The free edge of the cusp is pulled downwards. There is fibrous attachment of the cusp to the crest of the septum.

cusp to be pushed into the defect by the diastolic pressure.[7] This pulls the cusp away from the center of the aortic orifice and reduces the coaptation area. This increases the stress of the free edge of the cusp. The billowing of the deformed right coronary cusp is what is wrongly referred to as right coronary cusp prolapse (Fig 3).

2. Fibrous attachments progressively develop from the periphery of the defect, fixing the cusp and reducing its height (Fig 4).
3. Once the height of the cusp is reduced, a vicious circle is initiated; the apposition of the free edge of the right cusp occurs lower than the free edge of the two other cusps, limiting the coaptation surface. The stress on the free edge increases and elongation of the right free edge appears. Regurgitation becomes visible on echo.
4. The elongation of the right free edge increases progressively, the aortic regurgitation worsen. The regurgitation becomes significant enough to generate a secondary thickening and retraction of the cusp tissue and thickening of the free edge. The right cusp progressively acquires more mass and gains inertia. The fine mechanics of the aortic valve is permanently altered (Fig 5).
5. The regurgitation is creating secondary lesions to the two other cusps.

Indications

We offer this surgery to all patients with infundibular VSD in whom traces of aortic regurgitation appears or increases during follow-up. Infundibular VSD without aortic regurgitation and no deformation of the right aortic cusp should benefit from medical follow-up, especially if the VSD is very small

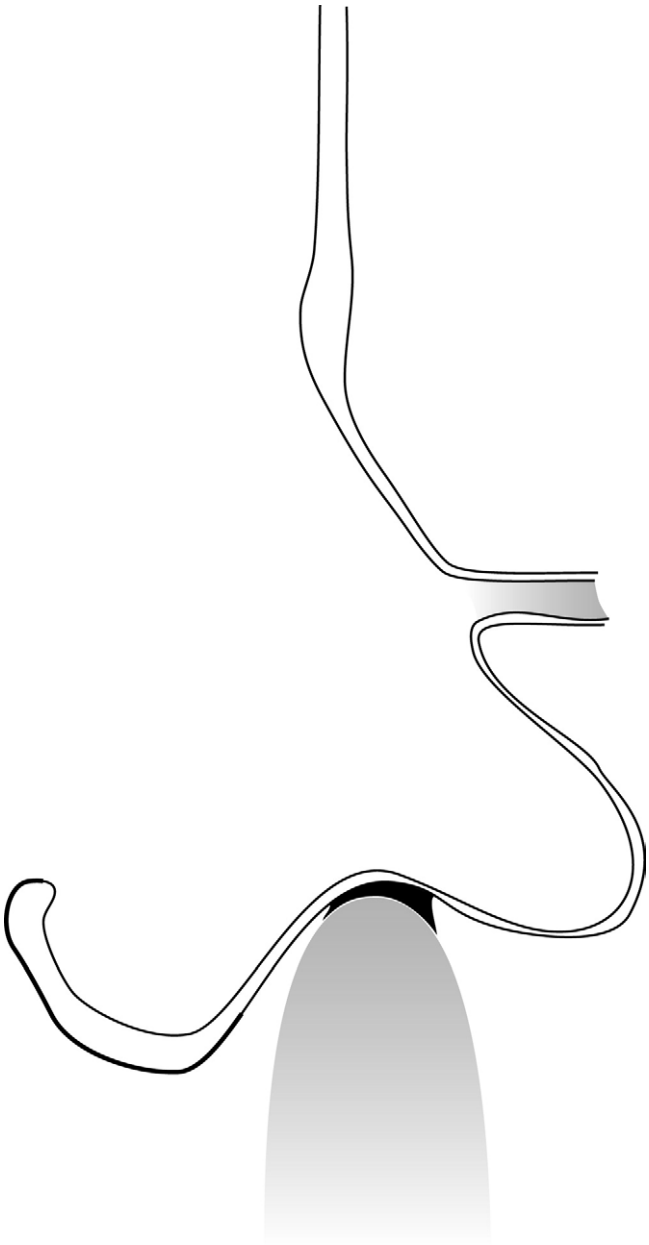

Figure 5 The height of the cusp is limited while the free edge is elongated. There are significant alterations of the cusp tissue.

(<5 mm).[6] There is no evidence that the operation on a patient with no aortic regurgitation will prevent the apparition of the aortic regurgitation. There are even suggestions that it does not.[8] On the other hand, we insist that patients with infundibular VSD and regurgitation greater than moderate should not be offered this valve repair. A satisfactory functional surface of apposition between the cusps cannot be restored if the secondary lesions to the cusps are severe. A valve replacement with either a Ross procedure, a mechanical prosthesis or an aortic valve repair with cusp extension of all three cusps should be performed instead, depending on the surgical preference and experience.[9]

Surgical Technique

The technique we use is directly derived from our understanding of the pathophysiology. The aim is to restore the normal height of the cusp by repositioning the hinge point of the right coronary cusp, hence a normal coaptation surface.

Cardiopulmonary bypass is established between the body of the right atrium and the ascending aorta. If an associated lesion needs to be addressed or the aortic regurgitation is the result of severe disturbance of the aortic valve anatomy, and a valve replacement with the autograft procedure may be indicated after inspection of the lesions, then a bicaval cannulation is indicated. Bypass is conducted at 32°C. The interatrial grove is slightly dissected inside the right superior pulmonary vein to expose some part of the septum. When full flow is reached, the cross clamp is applied and the left side cavities are vented through a stab incision in the interatrial septum in the space previously dissected. An oblique aortotomy extended into the noncoronary sinus is performed when the heart is emptied. Antegrade cardioplegia is directly delivered into the coronary ostia. Stays sutures are put in to allow for a large and steady exposure; this may include stays across the superior aspect of the commissures pulling cephalad. The valve and the defect are then inspected.

By lifting the nadir of the right coronary cusp, the defect is revealed. It is confined between the two commissures of the

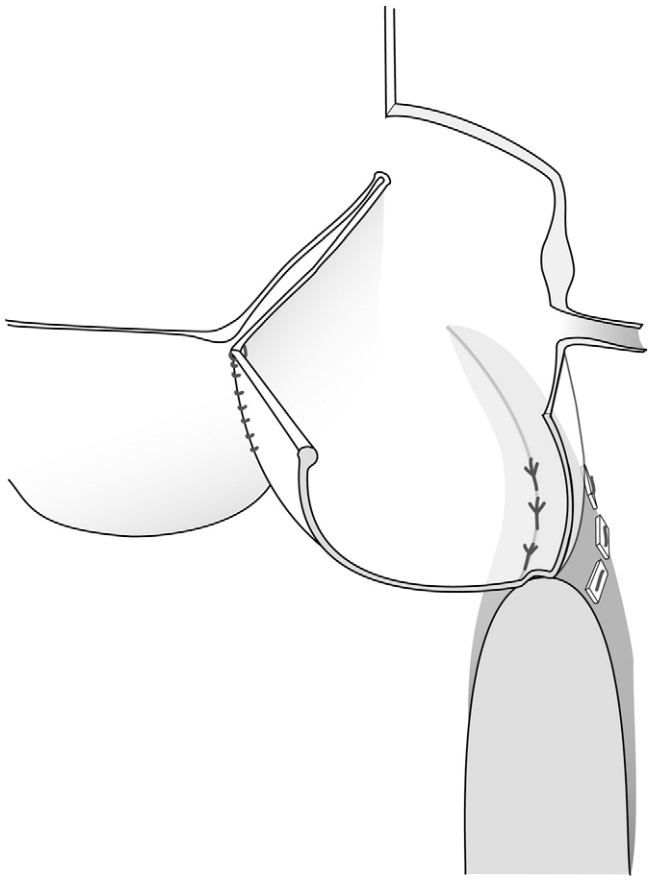

Figure 6 Repair after triangular resection of the excess of length of the free edge and reconstruction.

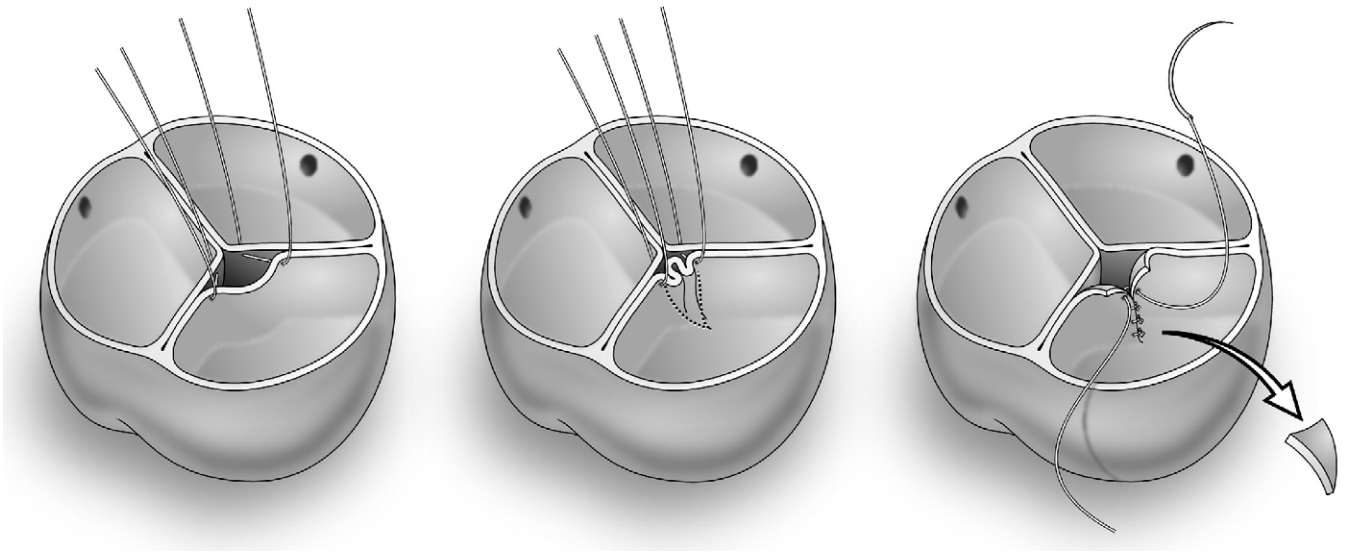

Figure 7 Shortening of the free edge with triangular resection.

right coronary cusp. It is larger than what would suggest the echocardiographic study or the importance of the shunt. The inspection will try to establish the precise site of the crest of the septum, the thickness and extension of the secondary fibrous attachment of the cusp to the crest of the septum toward each commissure. The inspection will differentiate this secondary lesion from the normal inter-leaflet triangles. Then the most important part of the inspection takes part: the inspection of the right coronary cusp itself. The amount of restriction to its height and the secondary elongation of the free edge are carefully appreciated. The height of the right coronary cusp is evaluated and compared with the height of the left coronary cusp or the non-coronary cusp. A template of thick silk is made and used to measure the height of the more normal of the latter two cusps. This template is compared with the height of the right coronary cusp. To evaluate the elongation of the free edge, again, the left and non-coronary cusps are used as benchmarks. A 6/0 stitch is transfixing the nodulus of Arantius of the left coronary cusp, as would a Frater apposition stitch. With stretching applied on the free edge of the right coronary cusp, it is opposed against the corresponding half of the left coronary cusp, the 6/0 stitch is transfixing the free edge of the right coronary cusp immediately at the point facing the nodulus of Arantius in the left coronary cusp. A symmetrical maneuver is performed with the free edge of the noncoronary cusp. When this is completed, the distance between the two sites of insertion of the suspension stitches correspond with the excess of free edge length of the right coronary cusp, if any.

The repair of the defect is part of the repair of the aortic valve. It uses a strict transaortic approach. To achieve this, the first step is to mobilize the cusp from the edge of the septum if it is fixed. All thickening of the cusp and secondary fibrous attachments are divided. This extends close to both commissures of the right coronary cusp. A series of mattress sutures with pledgets are then applied onto the right side of the septum. The entry points of the stitches are into the right side of the septum and the exit points precisely at the crest of the defect. The stitches transfix the cusp following a line that will become the new hinge point. The position of this line is set so as to restore the height of the right cusp identical to the one of the two facing counterparts and is defined with the silk template described previously. The mattress sutures are tied on the aortic side of the cusp. This is done directly if the sinus of Valsalva is mildly deformed or not at all (Fig 6). If the sinus of Valsalva is grossly deformed, then the mattress sutures are tied over a patch of autologous pericardium. The latter will be used to reduce the height of the sinus underneath the right coronary ostia.

The procedure is completed by a reduction of the free edge of the right coronary cusp if it is elongated. We use a triangular resection at the mid-portion of the free edge and direct reconstruction with interrupted 7/0 sutures (Fig 7). The amount of resection has been determined precisely by the evaluation of the excess of free edge length. The resection should be slightly less then the distance between the two marker stitches to allow for reconstruction. The height of the triangular resection should not be greater than one half of the height of the cusp to keep a nice depth to the cusp. We believe that a plication of the free edge at the commissure level according to the technique described by Trusler et al[10] is to be avoided because it compromises the commissure and can easily lead to overcorrection.

The procedure is monitored with pre and post bypass transesophageal echocardiographic study.

At an early stage of the development of this technique the procedure also included the insertion of a patch on the aortic aspect of the sinus of Valsalva, as described by Bonhoeffer et al,[3] when a severe deformation of the sinus was associated. As we moved away from this pathophysiologic interpretation, we believe that this patch is solely indicated in the presence of an aneurysm of the right sinus of Valsalva, which we have not encountered in our series. Incidentally, the patch may have the

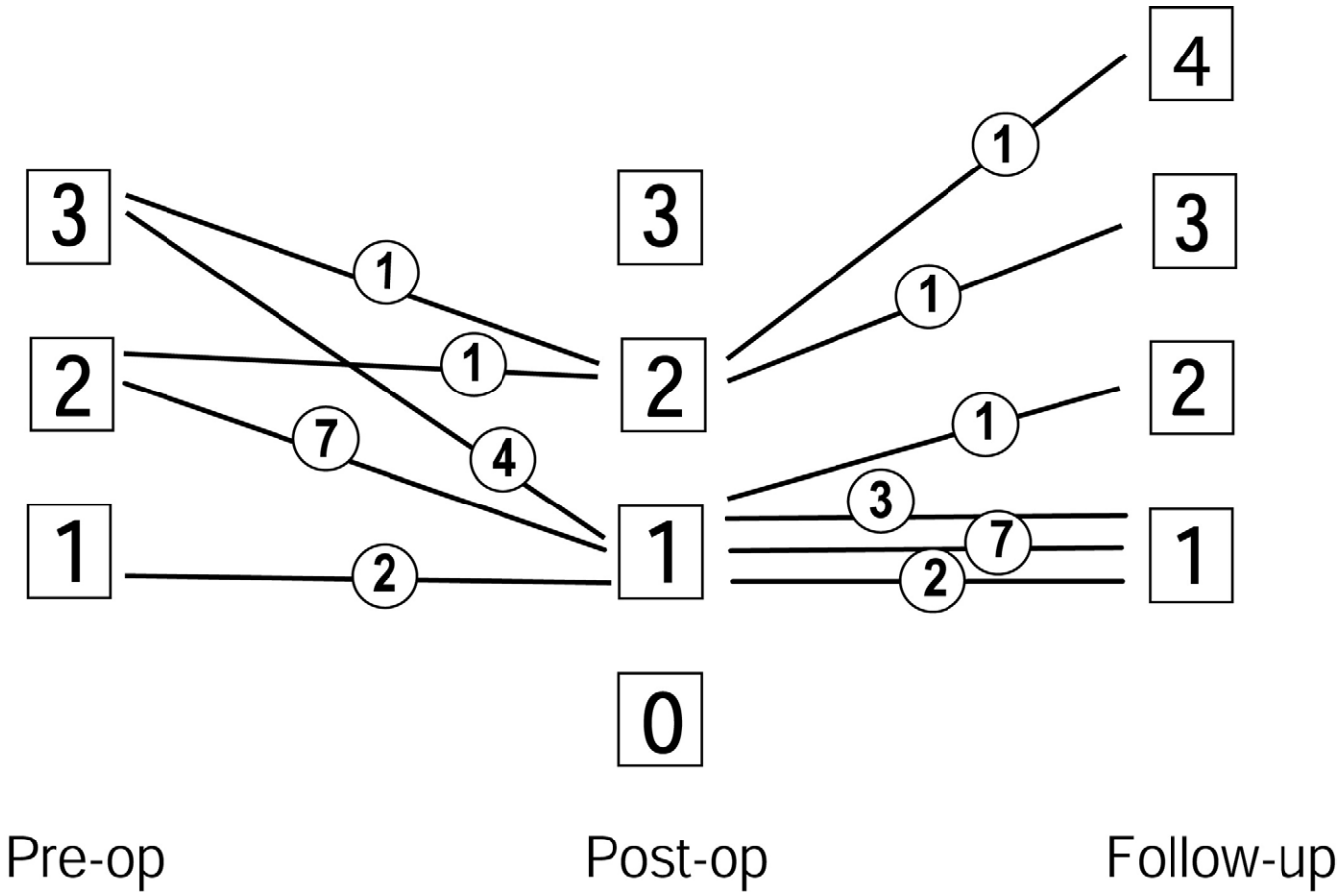

Figure 8 Evolution of the aortic regurgitation in the 15 patients.

benefit of protecting the aortic aspect of the cusp from the aggressive edges of the knots when polypropylene is used.

Clinical Experience

Between 1996 and 2005, 15 patients were operated on at the Royal Children's Hospital, Melbourne, using the above described technique. Median age at operation was 4.7 years (range, 1.3 to 19 years). There were four females. All patients had aortic regurgitation ranging form trivial (n = 2), mild (n = 8), and moderate (n = 5).

Six patients had a patch in the right sinus of Valsalva inserted. All patients had a simple apposition of the cusp to the crest of the septum after mobilization of the cusp. Seven patients had a triangular resection at the central portion of the free edge of the right coronary cusp.

The grade of the aortic regurgitation was proportional to the age of the patient; this reached statistical significance for mild (median, 4.1 years) and moderate (median, 8.6 years) (P = .02).

Follow-Up

Three overseas patients had short follow-up (less than 3 months). The other patients had a median follow-up of 14.5 months (range, 3 to 101 months) and recent echocardiographic study. There were no early deaths, but there was one late death. The patient was operated on at the age of 1 year and 3 months with apparition followed by rapid progression of the aortic regurgitation from the time of diagnosis in infancy. At the time of the surgery, the regurgitation was mild. The surgery was limited to the VSD closure as described with a pericardial patch to line the aortic aspect of the right sinus of Valsalva. No resection of the aortic cusp was performed. The immediate postoperative result was good with trivial regurgitation. The regurgitation increased to severe within 4 years. The patient was reoperated on electively and had an aortic valve repair with cusp extension of all three cusps with autologous pericardium treated with glutaraldehyde. The postoperative result was very satisfactory with no aortic regurgitation and good ventricular function. The patient had an arrest in the Medihotel on the sixth postoperative day. Resuscitation was not successful despite rapid deployment of left ventricular assist device. No signs of recurrence of aortic regurgitation were noted on the echocardiography study on left ventricular assist device.

All the other patients were fine at follow-up. Figure 8 shows the state of the aortic regurgitation at the time of discharge and at follow-up. One patient had moderate regurgitation at the time of repair. The result of the surgery was unsatisfactory and aortic regurgitation started to progress again after 1 year.

Comments

The results of this short series suggest several comments.

Indication to operate in patients with no aortic regurgitation and no right cusp deformation should be weighed carefully.

Patients who demonstrate progression of their aortic regurgitation will continue to do so without intervention.

The surgery is more efficient in reducing the aortic regurgitation when the latter is mild or mild to moderate at the most.

The result of the surgery is more stable in patients with initially little regurgitation.

The surgery never achieves perfect result because damages to the aortic cusp (thickness of the free edge, rigidity of the body) are irreversible.

References

1. Chauvaud S, Serraf A, Mihaileanu S, et al: Ventricular septal defect associated with aortic valve incompetence: Results of two surgical managements. Ann Thorac Surg 49:875-880, 1990
2. Yacoub M, Khan H, Stavri G, et al: Anatomic correction of the syndrome of prolapsing right coronary aortic cusp, dilatation of the sinus of Valsalva, and ventricular septal defect. J Thorac Cardiovasc Surg 113:253-261, 1997
3. Bonhoeffer P, Fabbrocini M, Lecompte Y, et al: Infundibular septal defect with severe aortic regurgitation: A new surgical approach. Ann Thorac Surg 53:851-853, 1992
4. Lun KS, Li H, Leung MP, et al: Analysis of indications for surgical closure of subarterial ventricular septal defect without associated aortic cusp prolapse and aortic regurgitation. Am J Cardiol 87:1266-1270, 2001
5. Momma K, Toyama K, Takao A, et al: Natural history of subarterial infundibular ventricular septal defect. Am Heart J 108:1312-1317, 1984
6. Tatsuno K, Konno S, Ando M, et al: Pathogenetic mechanisms of prolapsing aortic valve and aortic regurgitation associated with ventricular septal defect. Anatomical, angiographic, and surgical considerations. Circulation 48:1028-1037, 1973
7. Ando M, Takao A: Pathological anatomy of ventricular septal defect associated with aortic valve prolapse and regurgitation. Heart Vessels 2:117-126, 1986
8. Tomita H, Arakaki Y, Ono Y, et al: Severity indices of right coronary cusp prolapse and aortic regurgitation complicating ventricular septal defect in the outlet septum: Which defect should be closed? Circulation J 68:139-143, 2004
9. Schoof PH, Hazekamp MG, Huysmans HA: Pulmonary autograft in ventricular septal defect-aortic insufficiency complex. Ann Thorac Surg 61:1005-1006, 1996
10. Trusler GA, Moes CA, Kidd BS: Repair of ventricular septal defect with aortic insufficiency. J Thorac Cardiovasc Surg 66:394-403, 1973