# Left Ventricular Outflow Tract Anomalies

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**Introduction**

Congenital left ventricular outflow tract obstruction accounts for 10% of all congenital heart disease. For practical purposes, the site of obstruction is classified anatomically as valvar, subvalvar, or supravalvar, or as a combination of the three (multilevel stenosis).

The decision-making process and planning of operative care is complicated by the heterogeneous make-up of patients with left ventricular outflow tract obstruction. In neonates, the critical point is to decide whether biventricular repair is feasible. Patients with isolated stenosis and well-developed left ventricles are optimal candidates for biventricular repair. At the other end of the spectrum, a left ventricular outflow tract obstruction could be part of complex congenital cardiac malformations with a morphologically or functionally (endocardial fibroelastosis) borderline left ventricle, with multiple sequential outflow obstructions (Shone syndrome, hypoplastic left heart syndrome, etc.), when the single-ventricle pathway might be the optimal approach.

Growth, an active lifestyle with the appropriate activity level, and the difficulties in medical compliance represent another specific set of requirements posed by this subset of patients. In general, the prosthetic materials should not compromise cardiac growth and lifestyle. In particular, reconstruction of the aortic valve and pulmonary autograft procedure for replacement of the aortic valve and aortic root has dramatically changed the approach to children with congenital aortic valve disease and complex left ventricular outflow tract obstructions. On the other hand, in some children, the best alternative is to use a prosthetic valve or allograft, despite the well-known drawbacks of these procedures.

**Aortic Stenosis in Neonates and Infants**

**Anatomy**

A reduced cross-section area in critical aortic stenosis is the result of (1) deficiency in or absence of one or more commissures, leading to a unicuspid or bicuspid aortic valve; (2) immaturity with myxomatous change and thickening of the valve cusps, with or without commissural fusion; and (3) hypoplasia of the valvar annulus.
The unicuspid valve usually has an eccentric, pinhole orifice with one defined commissure or no commissural attachment lateral to the aortic wall. Even in the absence of commissural fusion, the valve is severely stenotic from birth. The dysplastic bicuspid or tricuspid aortic valves might have a myxomatous appearance and thick cusps with a variable degree of peripheral commissural fusion. This type of valve produces a dome with a central opening. It is believed that swollen cusps, rather than the fusion of commissures, cause the obstruction. The aortic annulus and ascending aorta are usually hypoplastic. In general, immaturity (gelatinous, myxomatous primitive tissue) and incomplete development (poorly defined cusps, bicuspid or unicuspid morphology, borderline aortic annulus) are typical features of critical neonatal aortic valve stenosis.

Indication for Surgery

Critically ill neonates should be stabilized by aggressive resuscitation, including mechanical ventilation, inotropic support, and correction of metabolic acidosis, while patency of the ductus is maintained by prostaglandins. A trial discontinuation of prostaglandins can be undertaken if the structures on the left side are well developed, left ventricular function is adequate, and the degree of aortic valve stenosis is not severe. No intervention is needed if ductal closure is tolerated. On the other hand, dependency on the ductal circulation is a clear indication for operation.

Approach and Cardiopulmonary Bypass Strategy

The heart is approached through a median sternotomy. After opening the sternum, the thymus is nearly completely removed, and a patch of autologous pericardium is harvested. The patch, pretreated in glutaraldehyde for 15 min, is used for reconstruction of the valve, if necessary. The operation is performed on cardiopulmonary bypass with normothermia or mild hypothermia (35–32°C). After systemic heparinization, the aorta is cannulated as high as possible to leave enough space for working on the ascending aorta. Two venous cannulas are used if additional defects (atrial septal defect) must be addressed. The left atrial vent is placed to keep the operating field bloodless. Antegrade delivery of crystalloid cardioplegia is used.
Open Valvotomy for Critical Aortic Stenosis in the Newborn

The Goal of Surgery
Open surgical valvotomy allows construction of a cusp anatomy that is as close as possible to the normal anatomy. Shaving of thickened cusps, excision of obstructive myxomatous nodularities, and mobilization of the cusps effectively increase the orifice area of the valve in any type of morphology, with a minimal risk of creating regurgitation, thus preserving the native aortic valve in the majority of patients in the long run.

Patient Characteristics

**Age at surgery:** 14 days  
**Diagnosis:**  
1. Critical aortic stenosis  
2. Unicuspid aortic valve  
3. Atrial septal defect  
**Procedure:**  
1. Surgical commissurotomy  
2. Reconstruction of the anterior commissure and creation of a bicuspid arrangement  
3. Shaving of cusps  
4. Resection of subvalvar nodules  
5. Direct closure of the atrial septal defect  
**History:**  
The patient presented with severe aortic stenosis since birth, however, with good function of the left ventricle.

Specific Steps of Surgery

**DVD Clip 1**
Preoperative echocardiogram.
An oblique, inverted–hockey stick–type of incision is made from the anterior aspect of the aorta towards the noncoronary sinus of Valsalva.

The morphology of the valve, the arrangement of the commissures, and localization of the false raphe and orifices of the coronaries are noted. The degree of peripheral commissural fusion and nodular excrescences on the cusps composed of myxomatous tissue and the thickened part of the cusps are identified.
The fused commissures are carefully divided with a knife, ensuring that the leaflets are well supported and not liable to prolapse. The false raphe, if present, does not provide any lateral support for the cusp, and it should not be incised. Obstructive myxomatous and fibrous nodules on the cusps are removed. This implies careful and meticulous thinning/shaving of the cusps with a knife.

The opening of the valve, the mobility of the cusps and the geometry is assessed. Correctly performed “shaving” greatly improves the mobility of the cusps and the effective orifice area of the aortic valve. The effective orifice area is checked by “olive” (an olive-shaped probe is inserted into the orifice), and the commissurotomy is extended, if necessary. Even a short commissurotomy incision could result in adequate opening of the outflow orifice, because the flow is a function of the fourth power of the radius.
The aortotomy is closed with a running suture. After rewarming, the patient is weaned from the cardiopulmonary bypass. Transesophageal echocardiographic assessment of the left ventricular function, aortic valve function, and flow characteristic in the left ventricular outflow tract should be carried out routinely. The ductus arteriosus should always be ligated if the patient is stable. If cardiac output is not adequate, even on aggressive inotropic support, the duct can be left opened and maintained patent on prostaglandins to provide additional systemic perfusion from the right ventricle.
Aortic Stenosis in Children

Anatomy

Although all anatomic variations are possible, the valve is bicuspid in about 70% of cases, with the two commissures arranged as anterior and posterior, or left and right. There may be a third raphe or false commissure. There are usually variable degrees of peripheral fusion of one or both commissures, creating a stenosis. Stenosis can occur without commissural fusion, simply because of thickened or myxomatous cusps and a bicuspid configuration. If the free edges of both thickened bicuspid cusps are taut with no extra length and equal in length to the diameter of the aortic root, they cannot open completely and thereby produce obstruction. The aortic annulus is usually of adequate size. Abnormal tricuspid aortic valves might not be obstructive during early infancy, but could become stenotic later in life due to cusp thickening and calcification.

Indication for Surgery

Surgery for aortic valve stenosis is indicated in symptomatic patients when the peak Doppler gradient exceeds 40 mmHg and left ventricular hypertrophy and echocardiogram changes are notable. In asymptomatic patients, a peak gradient more than 60 mmHg is an indication for procedure. Although each patient should be evaluated individually, evidence suggests that early intervention, before the left ventricle develops myocardial changes, might be beneficial. Currently, intervention/surgery is indicated even in asymptomatic patients with a peak gradient between 40 and 60 mmHg, in order to promote aortic annulus growth.

Approach and Cardiopulmonary Bypass Strategy

The heart is approached through a median sternotomy. The standard technique of cardiopulmonary bypass with mild hypothermia (32°C) is used. Antegrade delivery of cardioplegia is performed. A left ventricular vent is inserted through the entrance of the right pulmonary veins.
The management of the operation is similar to that described for neonates. Visualization of the aortic valve anatomy is much easier. In order to maximize the effective orifice area, especially in a bicuspid scenario, the commissurotomy can be extended in circumferential fashion beyond the commissure into the aortic wall, splitting the aortic wall into two layers. If completion of the commissurotomy does not result in adequate opening, despite the normal annulus dimension, conversion to a three-cusp valve can be considered. Usually there is a prominent, thickened, even calcified, false raphe. Under these conditions the false raphe is incised. Thickened and immobile parts of the newly created cusps are completely removed, leaving as much native mobile tissue as possible. Autologous or bovine pericardial augmentation of these two cusps is performed. Augmentation of the third cusp depends on the effectiveness of coaptation with the already augmented cusps. Usually, the symmetric augmentation of all the cusps involved provides sufficient overlapping of the cusps and optimizes the height and width of the intercusp triangle, which governs the area of cusp apposition.

Alternatively, the raphe between the other unsupported cusps is incised, and a pericardial triangle patch is folded and sutured along both edges of the divided raphe and vertically to the aortic wall to provide support in the diastole, preventing cusp prolapse. The supravalvar aortic region is augmented as part of the aortotomy closure if there is a risk of narrowing. Should there be poststenotic dilatation of the ascending aorta, wedge resection of the anterior part of aorta is performed to restore an optimal sinotubular dimension.

Aortic Insufficiency in Children

Anatomy

The morphology depends on the etiology. In children, aortic regurgitation is associated with certain congenital heart diseases (ventricular septal defect, truncus arteriosus, tetralogy of Fallot, subaortic membrane, etc.) or with bicuspid aortic valves with or without coarctation. Currently, post-interventionally induced aortic regurgitation with residual aortic stenosis is frequently seen.
Predominant morphological factors contributing to aortic regurgitation after balloon dilation of the aortic valve are a combination of anterior commissural avulsion, cusp dehiscence with retraction (presumably caused by a long-standing, balloon-induced tear), cusp tear or perforation, central incompetence due to calcified cusps and sinus of Valsalva dilation, deficient cusps, and free–cusp–pal edge adhesion to the aortic wall. Annulo-aortic ectasia can be the result of dilatation of the aortic root due to tissue disorder (Marfan syndrome, Loeys–Dietz syndrome, Ehlers–Danlos, Turner syndrome). This process begins in the sinuses of Valsalva, with pear-shaped aneurysmal enlargement of the aortic root, creating valvular incompetence by dilatation at the commissural level. Rheumatic processes are another cause of aortic regurgitation, with shortening of the cusps between their free edges and the annular attachments. This results in rolling of the free edges and central incompetence. Less common courses are bacterial endocarditis, myxomatous degeneration, spontaneous cusp rupture, etc. Pure aortic regurgitation instead of insufficiency is very rare.

**Indication for Surgery**

The timing of surgery in patients with chronic aortic regurgitation is controversial. Accepted indications are the presence of symptoms, increasing significant left ventricular dilation with or without dysfunction, or both, in the setting of moderate or more severe aortic regurgitation. In order to achieve complete recovery of the left ventricular function, the surgery should be performed before the z-score of the preoperative left ventricular end-diastolic dimension is no greater than +4.

**Approach and Cardiopulmonary Bypass Strategy**

The heart is approached through a median sternotomy. The standard technique of cardiopulmonary bypass with mild hypothermia (32°C) is used. Due to aortic regurgitation, the cardioplegia is delivered directly to the coronaries. If a longer aortic cross-clamp time is expected, retrograde delivery of cardioplegia should be considered. A left ventricular vent is inserted through the entrance of the right pulmonary veins.
The Goal of Surgery

In normal hearts, the aortic root, including the flexible sinotubular junction, expands during systole, straightening the cusp-free edges and maintaining a constant strain in order to minimize fatigue stress. Apart from this mechanism, proper coaptation depends on the aortic annulus, the sinotubular dimension, and the height and position of each commissure in relation to the other commissures. Commissural attachments should be always higher than the plane of the valve, giving the cusps the necessary depth for coaptation. The crescent-shaped portion of the cusp’s free edge forms the area of coaptation. Abnormalities in the length or structure of the free edges result in poor mobility or poor coaptation. A deficient cusp usually results from a long-standing, balloon-induced tear with retraction of the free cusp edge. Anatomic elements necessary for valve repair include a sufficient annulus diameter, mobile cusps, or cusps that can be mobilized by resection or shaving off any excess fibrous tissue, and an ability to achieve coaptation without inducing stenosis.

The goal of reconstruction is to restore the normal geometry of the valve, leaving as much as possible of the native valve and tailoring the construction to individual cusps. The optimal patch material is autologous pericardium, pretreated in glutaraldehyde, with excellent mechanical and handling properties and with a diminished rate of calcification, retraction, and fibrosis. The fixation time of 15–30 min improves the handling and increases the strength of the pericardial patch. The other alternative is bovine pericardium. This material is highly resistant to calcification, supports the growth of host cells, has excellent handling characteristics, and does not require a pre-implantation rinse, making it rapidly available.

The first step of the operation is to relieve commissural fusion and extensive thinning of the valve cusps. Then, the appropriate technique of repair for aortic valve regurgitation is chosen based on the underlying pathology. There are several scenarios:

1. Perforations or tears in the aortic cusps are repaired either with a direct suture or with pericardial patches.
2. Redundant tissue is corrected by shortening the coaptation edge to match the adjacent cusps. For the assessment of cusp prolapse in a tricuspid anatomy, a fine suture is passed through the noduli of Arantius and placed under trac-
tion to estimate the relative length of all free cusp margins. In bicuspid aortic valves, radial tension is exercised on the opposite commissures to identify the extend and the location of prolapse. A redundant, prolapsing cusp can be repaired by several similar methods:

a) Shortening is provided by triangular resection of redundant tissue. A small, extra amount of tissue is left for incorporation into the suture line. The resected margins are approximated by means of interrupted or continuous polypropylene sutures.

b) Plication of redundant tissue in the prolapsed cusp achieves the same effect. No resection of valve tissue is needed.

c) The redundant portion of the cusp can be plicated and secured against the aortic wall with a 5-0 mattress stitch supported with pledgets.

d) Correction of the cusp’s length does not automatically result in the correction of the cusp prolapse. To improve coaptation of the cusps, the free-edge reinforcement of the leaflet can be applied, reducing the length of the cusp-free margin. Free-margin reinforcement is performed by an over-and-over running GORE-TEX® suture. Both ends of the suture are suspended at the level of the corresponding commissures, reinforcing and shortening the free edge.

3. More extensive cusp destruction, with deficient cusps, requires an extension procedure using a pericardial patch. The length of the half-moon–shaped patch should be 15% greater than the diameter of the aorta. The height is governed by the extent of augmentation and anticipated height of the newly created commissure. The destroyed part of the cusp is removed, and the patch is tailored to achieve a “normal” morphology of the newly created cusp. A deliberate overcorrection in the cusp reconstruction is attempted to provide additional material to extend the line of coaptation of the cusps 3–5 mm above the line of original coaptation and 2–3 mm above the sinotubular bar. The running 6-0 monofilament suture line is carried from the base of the native cusp to each commissure, then up onto the aortic wall, 2–3 mm above the original commissural insertion, avoiding the coronary ostia. With each patch in place, the newly constructed pericardial cusp edges are trimmed to provide a uniform cusp depth. The commissures are suspended and reinforced with 5-0 monofilament sutures. The small ascending aorta should be enlarged with a patch, especially if a three-cusp extension is performed, to avoid stenosis at the sinotubular junction.
4. Mild dilatation of the annulus can be managed with a subcommissural annuloplasty. This reduces the circumference of the annulus and increases cusp coaptation, without interfering with the range of motion of the cusps. The amount of plication depends on the level at which the sutures are placed. The lower the sutures are placed at the commissures, the greater the annular plication and degree of cusp coaptation. Horizontal mattress sutures with pledgets are placed through the annulus on each side of the commissure and tied. Care is taken to avoid contact with the cusps and coronary ostium.

5. For important root dilatation with progressing aortic valve insufficiency due to tissue disorder (Marfan syndrome, Ehlers–Danlos, Turner syndrome, etc.) or congenital heart diseases (bicuspid aortic valve), a re-implantation valve-sparing procedure should be used. The diseased root and all three sinuses of Valsalva are excised, and the valve is preserved. The tubular ascending aortic graft with the prefabricated sinuses is firmly anchored proximally at the ventriculooaortic junction below the leaflets, with the commissures sewn inside a Dacron graft. The prefabricated sinus of Valsalva might limit the potential for leaflet damage. The reimplantation technique minimizes the risk of late aortic annulus dilatation, which is seen in patients after the remodeling procedure.

6. In the case of central cusp incompetence with dilatation of the noncoronary sinus of Valsalva, a reduction plasty is performed to reduce commissural splaying. A wedge of the noncoronary sinus wall is resected, followed by primary closure of the aortotomy.

7. In isolated cusp prolapse associated with normal root anatomy, a sinotubular junction plasty is performed to improve coaptation of the leaflets. Pledged 4-0 monofilament sutures are placed in the middle part of the intercommissural segment. The dilated sinotubular junction or the ascending aorta, or both, might require plication, replacement with prosthesis, or resection.

Intraoperative assessment of the reconstruction of the valve is essential. The effective orifice area of the valve should always be checked by dilators. A dimension of 1–2 mm larger than that based on z-score is necessary to achieve a minimal gradient. After weaning, a direct measurement of the pressure gradient
between the left ventricle and ascending aorta should be obtained. The trans-esophageal echocardiogram is an essential tool for assessment of the function of the left ventricle, of gradients, and of the insufficiency of the aortic valve. If the peak gradient is >40 mmHg and aortic insufficiency is moderate or more, attempts to reduce these residuals should be considered. In general, residual stenosis is better tolerated than insufficiency.

The complex underlying anatomy requires the use of a combination of the techniques described above to achieve a desirable result.

**Subcommissural Downsizing of the Aortic Annulus, with Asymmetric Resection of the Sinus Valsalva**

*Patient Characteristics*

<table>
<thead>
<tr>
<th>Age at operation:</th>
<th>1 year</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Diagnosis:</strong></td>
<td></td>
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<tr>
<td>1. Severe central aortic regurgitation due to dilatation of the aortic root</td>
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<tr>
<td>2. Aneurysm of the noncoronary sinus of Valsalva</td>
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<tr>
<th><strong>History:</strong></th>
<th>Failure to thrive</th>
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<tr>
<th><strong>Procedure:</strong></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Subcommissural downsizing of the aortic annulus</td>
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</tr>
<tr>
<td>2. Downsizing of the noncoronary sinus of Valsalva</td>
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</tbody>
</table>

**Specific Steps of Surgery**

The operative finding corresponded (Fig. 3.1) with the preoperative diagnosis of root dilatation, with aneurysm of the noncoronary sinus of Valsalva. The subcommissural annuloplasty was performed first (Fig. 3.2) to reduce the circumference of the annulus and increase cusp coaptation without interfering with the range of motion of the cusps. Subsequently, the wedge of the noncoronary sinus wall was resected (Fig. 3.2) to reduce commissural splaying, followed by primary closure of the aortotomy (Fig. 3.3).
Fig. 3.1

Fig. 3.2

Fig. 3.3
Resection of the Subaortic Membrane and Pericardial Patch Augmentation of the Aortic Valve

Patient Characteristics

Age at operation: 15 years
Diagnosis:
Severe aortic regurgitation with subaortic membrane
History:
Progressive combination of aortic regurgitation and subvalvar stenosis

Procedure:
1. Resection of the subaortic membrane
2. Commissurotomy and shaving of the cusps
3. Pericardial patch enlargement of all three cusps of the aortic valve

Specific Steps of Surgery

After opening of the aorta, a circular membrane (Fig. 3.4) and a tricuspid, thickened aortic valve with partial fusion of commissure between the left and right cusps was found (Fig. 3.5).
The membrane was first completely removed (Fig. 3.6); this was followed by shaving of the valve and commissurotomy (Fig. 3.7). The patches, made of autologous pericardium, were tailored to achieve a “normal” morphology of the newly created cusps. Deliberate overcorrection was attempted in the reconstruction of the cusps, to provide additional material to extend the line of coaptation (Figs. 3.8, 3.9).
Aortic Valve Reconstruction with Pericardial Extensions, Resection of the Subaortic Membrane, and Closure of the Aneurysm of the Sinus Valsalva

Patient Characteristics

**Age at operation:** 13 years

**Diagnosis:**
1. Severe aortic regurgitation
2. Moderate subvalvar and valvar stenosis
3. Aneurysm of the left sinus Valsalva
4. Status post–bacterial endocarditis

**History:**
1. At the age of 10 years, the patient suffered from bacterial endocarditis (*Staphylococcus aureus*).

2. Due to the symptoms (syncopy), progressive aortic regurgitation, and dilatation of the left ventricle, an operation was indicated.

**Procedure:**
1. Resection of the subaortic membrane
2. Direct closure of the aneurysm of the sinus Valsalva
3. Commissurotomy and shaving of the cusps
4. Pericardial patch enlargement of all three cusps of the aortic valve
Specific Steps of Surgery

**Clip 1**
Preoperative findings.

**Clip 2**
Opening of the aorta.

**Clip 3**
Resection of the subaortic membrane.
Closure of the aneurysm of the sinus Valsalva.

Assessment of the anatomy and function of the valve.
Augmentation of left and right coronary cusps with autologous pericardium.

Postoperative echocardiogram.
Tricuspidalization of the Unicuspid Aortic Valve – Reconstruction of Two Cusps with the Native Tissue and Creation of the Remaining Cusp Using Autologous Pericardium

Patient Characteristics

**Age at operation:** 14.5 years

**Diagnosis:**
1. Congenital aortic stenosis
2. Unicuspid aortic valve
3. Status post-ballooning
4. Severe aortic regurgitation

**History:**
1. At the age of 9 years, the patient underwent ballooning for severe aortic stenosis (pressure gradient > 120 mmHg), with subsequent development of aortic regurgitation.
2. Due to the symptoms, progressive aortic regurgitation and dilatation of the left ventricle, an operation was indicated.

**Procedure:**
1. Tricuspidalization of the unicuspid valve
2. Reconstruction of two cusps from native tissue
3. Creation of a third cusp using autologous pericardium

Specific Steps of Surgery

**DVD** Clip 1

Preoperative findings.
Tricuspidalization of the valve. The left coronary cusp and a new commissural post is fashioned. The noncoronary cusp is used as a reference point to adjust the length of the left cusp with the aim of achieving a symmetric arrangement.

The right coronary cusp is created from a patch of autologous pericardium. The length of the half-moon–shaped patch should be 15% greater than the diameter of the aorta. The 15% additional length accounts for a reduction in the pericardial cusp free-edge width that results from a purse-string effect with a running polypropylene suture. The height is governed by the extent of augmentation and anticipated height of the newly created commissure.
Reinforcement of the commissures.
Reconstruction of an Aortic Valve Destroyed by Bacterial Endocarditis – Closure of an Abscess Formation Invading the Annulus of the Aortic Valve and Anterior Leaflet of the Mitral Valve

Patient Characteristics

**Age at operation:** 19 years

**Diagnosis:**
1. Congenital aortic stenosis
2. Status post-ballooning
3. Severe aortic regurgitation
4. Aneurysm of the ascending aorta
5. Membranous subaortic stenosis
6. Status post-resection of the subaortic membrane, reconstruction of the aortic valve with pericardial extensions, and replacement of the ascending aorta with a prosthesis
7. Endocarditis of the aortic and mitral valves
8. Septic embolization to the brain with subsequent deafness on the left side

**History:**
1. At the age of 8 years, the patient underwent ballooning for severe aortic stenosis, with subsequent development of aortic regurgitation.
2. Due to symptoms at the age of 16 years, progressive aortic regurgitation with subaortic membrane, dilatation of the left ventricle, and development of an aneurysm of ascending aorta, the patient was indicated for reconstructive surgery. During the operation the membrane was resected, the aortic cusps were shaved and reconstructed using autologous pericardial extensions; the noncoronary sinus of Valsalva was downsized, and the ascending aorta was replaced with a prosthesis (Figs. 3.10–3.12). After surgery, there was trivial aortic regurgitation and no obstruction of the left ventricular outflow tract.
3. At the age of 19 years, the patient was urgently admitted with bacterial endocarditis (S. aureus), and septic embolization to the brain. After a short antibiotic treatment, she underwent reconstructive surgery.

**Procedure:**
1. Resection of the noncoronary cusp and debridement of the remaining cusps
2. Direct closure of the abscess formation
3. Direct closure of the perforation of the anterior leaflet of the mitral valve
4. Pericardial patch enlargement of the noncoronary cusp
Specific Steps of Surgery

Closure of the abscess formation and reconstruction of the noncoronary cusp.
Ross Operation

The Goal of Surgery
The aim is replacement of a stenotic or regurgitant aortic valve with an autologous pulmonary valve, which has growth potential, superior hemodynamic performance, and no need for anticoagulant therapy. The Ross procedure even allows replacement of the aortic valve earlier in life, thus avoiding repeated surgical reinterventions, which provide only short-term palliation and potentially exacerbate ventricular function. In the case of severe abnormalities or bicuspid anatomy of the pulmonary autograft, or a significant mismatch between aortic and pulmonary annuli in favor of the aortic annulus, one should consider abandoning the Ross operation.

Patient Characteristics

<table>
<thead>
<tr>
<th>Age at operation:</th>
<th>11 years</th>
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<tbody>
<tr>
<td>Diagnosis:</td>
<td>1. Shone syndrome</td>
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<td></td>
<td>2. Aortic stenosis</td>
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<td></td>
<td>3. Parachute mitral valve</td>
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<td></td>
<td>4. Hypoplasia of the aortic arch with coarctation</td>
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<tr>
<td>History:</td>
<td>1. Aortic valvotomy and repair of the arch at the age of 6 weeks</td>
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<td></td>
<td>2. Aortic reconstruction at the age of 8 months</td>
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<td>3. Gradual progression of aortic stenosis</td>
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<td>Procedure:</td>
<td>1. Ross procedure using a technique of complete root replacement</td>
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<tr>
<td></td>
<td>2. Reconstruction of the right ventricular outflow tract with pulmonary homograft</td>
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</tbody>
</table>

Specific Steps of Surgery
In children, a standard technique of complete root replacement is used. Extensive dissection and mobilization of the ascending aorta, the pulmonary trunk, and both pulmonary arteries are carried out. After aortic cross-clamping, the aorta is partially transected and the feasibility of valve repair is assessed. If the decision is made to proceed with the Ross operation, the main pulmonary artery is transected just proximally to the bifurcation of the pulmonary artery. The pulmonary valve is inspected to ensure no abnormality exists.
Preoperative findings.

The right and left coronary buttons are formed and dissected for maximal mobilization.

The aortic cusps and sinus wall are removed, leaving an approximately 3–5 mm cuff of the aortic wall in place.
The pulmonary autograft is harvested. The main pulmonary artery has been divided proximal to the branches, and the proximal vessel with the valve is harvested from the right ventricle. A circumferential muscle bar is taken with the graft. After harvesting the pulmonary valve, cardioplegia is delivered, and any bleeding points from the area of previous dissection are controlled by either diathermy or are over-sewn with a shallow suture.

In adolescents, in the case of a geometric mismatch of at least 3 mm in favor of the aortic annulus, the commissures on either side of the noncoronary sinus are plicated down with pledget-supported sutures to ensure the proper autograft fit and function. The pulmonary autograft is then sutured to the left ventricular outflow tract opening using a continuous suture.
Reinforcement of the seated autograft is performed.

The left coronary artery is reimplanted into the neoaortic root.
The neoaortic reconstruction is completed by proximal anastomosis with the ascending aorta. Before the anastomosis is completed, the right anterior commissure is marked on the outside of the autograft to avoid inadvertent injury to the valve leaflet during the implantation of the right coronary artery.

The right coronary artery is reimplanted into the neoaortic root.
A homograft or bovine jugular vein conduit is used for reconstruction of the right ventricular outflow tract.

A pulmonary valve homograft is anastomosed with the right ventricular outflow tract.
After de-airing, the aortic clamp is released and the patient is weaned from bypass. An echocardiogram should be routinely performed after weaning the patient from bypass.
Subvalvar Aortic Stenosis

Introduction

Subvalvar stenosis is an obstruction of the left ventricular outflow tract below the aortic valve. The most common form is a fixed obstruction due to a discrete membrane or a diffuse, tunnel-like obstruction. This condition occurs in patients with associated congenital heart diseases, which include ventricular septal defect, coarctation of the aorta, interrupted aortic arch, or atrioventricular canal, among others. Lesions can develop in patients with these defects before surgery, but can also appear and progress significantly after surgical correction of the associated defects. In addition, the discrete membrane can present as a primary defect. Other rare causes of subvalvar obstruction are abnormal attachments of the mitral valve, accessory tissue, abnormal insertion of the mitral papillary muscle, abnormal muscular bands within the left ventricular outflow tract, and space-occupying lesions in the left ventricular outflow tract, etc. Shone and associates described a complex of subvalvar aortic stenosis in association with a supravalvar mitral ring, parachute mitral valve, and coarctation of the aorta. Rarely seen hypertrophic obstructive cardiomyopathy usually creates dynamic subvalvar obstruction due to opposition of the anterior leaflet of mitral valve against the hypertrophied ventricular septum.

Anatomy

Fixed subaortic obstruction can be discrete or diffuse. The discrete form represents about 70% of cases, and it is characterized by a fibromuscular shelf located at the area of aortomitral continuity, with the extension toward the interventricular septum.

It is typically seen in patients with a more acute angle between the long axis of the left ventricle and the aorta. This angulation leads to imbalance in shear forces, which causes the proliferation of the tissue that forms the membrane. There could be an abnormal anatomic relationship with the mitral valve, which is displaced anteriorly, and it rotates in a clockwise direction.

Diffuse subvalvar aortic stenosis is caused by a circumferential narrowing that commences at the annular level and extends downward 1 to 3 cm into the
left ventricular cavity. It is often a secondary lesion seen after previous resection of a discrete subvalvar aortic membrane. Scarring from the initial resection in conjunction with an abnormally shaped left ventricular outflow tract could result in progressive fibromuscular proliferation and creation of a left ventricular outflow tract tunnel.

Hypertrophic obstructive cardiomyopathy is characterized by primary asymmetric ventricular septal hypertrophy, and a dynamic obstruction associated with abnormal systolic anterior motion of the anterior leaflet of the mitral valve. The histological hallmark of this disease is the presence of disorganized and bizarrely shaped hypertrophied myocytes.

**Indication for Surgery**

Symptoms associated with left ventricular outflow tract obstruction (syncope, angina, diminished exercise tolerance, etc.) are indications for surgery. Surgical intervention is indicated when the peak Doppler gradient across the left ventricular outflow tract is ≥30 mmHg in the discrete subvalvar aortic membrane and >60 mmHg in the tunnel-type obstruction. However, earlier indication for surgery, especially in infants, might prevent the development of endocardial fibroelastosis, thus preserving the mitral valve and left ventricular function. Surgery is also advocated in infants and children in the presence of aortic regurgitation, even when there is no significant gradient. Patients with a peak gradient <30 mmHg and no significant left ventricular hypertrophy are followed closely for progression, especially in the first several years of life. Indication for operation in hypertrophic obstructive cardiomyopathy is driven by the progression of symptoms despite the maximum medical therapy. In symptomatic patients, a peak Doppler gradient >60 mmHg under resting conditions or with provocation, preferably using physiological stress, is an indication for surgery. In asymptomatic patients, a peak Doppler gradient >75–100 mmHg at rest is an indication for surgery.
**Approach and Cardiopulmonary Bypass Strategy**

The heart is approached through a median sternotomy. The standard technique of cardiopulmonary bypass, including bicaval cannulation and mild hypothermia (32°C), is used. If long cardioplegic arrest of the heart is expected, myocardial protection is provided by retrograde cardioplegia. A left ventricular vent is inserted through the entrance of the right pulmonary veins.

**Resection of a Discrete Subaortic Membrane**

**The Goal of Surgery**

The circumferential excision of the fibrous ridge with septal myectomy is safe and provides efficient relief of obstruction. Aggressive resection of all structures causing flow turbulence and removal of pathological tissue from the valve leaflets might prevent development or progression of aortic regurgitation and might eliminate the substrate for recurrent obstruction. Nevertheless, recurrence remains a problem, especially in the presence of a predisposing associated congenital heart defect.

**Patient Characteristics**

<table>
<thead>
<tr>
<th>Age at operation: 7 years</th>
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<tbody>
<tr>
<td>Diagnosis:</td>
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<tr>
<td>1. Incomplete atrio–ventricular canal defect</td>
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<tr>
<td>2. Status post complete correction with a single-patch technique</td>
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<tr>
<td>3. Subaortic membrane with abnormal attachment of the mitral valve and prominent secondary chordae attached to the outflow tract</td>
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<tr>
<td>History:</td>
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<tr>
<td>1. At the age of 3 years, correction of incomplete atrio–ventricular canal defect</td>
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<tr>
<td>a. Adequate development and growth: trivial mitral regurgitation with no other residual defects</td>
</tr>
<tr>
<td>2. Gradual development of subaortic membrane and trivial aortic regurgitation</td>
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</table>

**Procedure:**

1. Resection of membrane
2. Myectomy of the left ventricular outflow tract
3. Resection of secondary chordae of the mitral valve attached to the outflow tract
Specific Steps of Surgery

**Clip1**

Preoperative echocardiogram

**Clip2**

Opening of the aorta and assessment of the underlying anatomy.
Resection of the membrane.

Resection of the secondary chordae of the mitral valve and myectomy.
Ross–Konno Operation with Resection of Endocardial Fibroelastosis in the Infant

The Goal of Surgery
The Ross–Konno operation is the procedure of choice in children with a multilevel type of left ventricular outflow tract obstruction and severe anomaly of the aortic valve. The reconstructed left ventricular outflow tract has growth
potential, and enlargement of the aortic annulus parallels somatic growth. The procedure allows easy access to the left ventricular myocardium for resection of endocardial fibroelastosis, normalizing the left ventricular outflow tract and the long axis of the left ventricle. The operation can be performed earlier in life, thus being successfully utilized for neonates or infants with critical aortic stenosis after failed first palliation (open valvotomy or ballooning) or for neonates with interruption of the aortic arch. The nature of the operation places two valves at risk for single-valve disease. However, many pediatric patients are not candidates for a Konno operation due to anatomic considerations and the lack of readily available, appropriately sized prostheses.

**Patient Characteristics**

**Age at operation:** 7 months

**Diagnosis:**
1. Severe left ventricular outflow tract obstruction with progressive aortic stenosis and mild aortic regurgitation
2. Extensive endocardial fibroelastosis effecting the papillary muscle
3. Hammock’s mitral valve with stenosis and progressive mitral regurgitation

**History:**
1. 1 Month of age: ballooning of aortic stenosis
2. Progressive aortic regurgitation with stenosis and mitral valve stenosis
3. 6 Months of age
   a. Aortic valve reconstruction with commissurotomy and pericardial patch extension of destroyed cusps after ballooning
   b. Splitting of the heads of the papillary muscle of the mitral valve
4. Progressive endocardial fibroelastosis with development of mitral regurgitation; mild aortic stenosis and regurgitation
5. Failure to thrive
6. 7 Months of age
   a. Ross–Konno operation
   b. Mitral valve plasty
   c. Resection of endocardial fibroelastosis
7. Progressive mitral regurgitation
8. 8 Months of age: mitral valve replacement

**Procedure:**
1. Ross–Konno operation
2. Mitral valve plasty
3. Resection of endocardial fibroelastosis
Specific Steps of Surgery

**Clip 1**

External anatomy of the heart.

**Clip 2**

In children, a technique of complete root replacement with anterior aortoventriculoplasty is used. After aortic cross-clamping, the aorta is partially transected, and the feasibility of valve repair is assessed. The main pulmonary artery is transected just proximal to the bifurcation of the PA.
After complete transection of the aorta, the right and left coronary buttons are formed and dissected for maximal mobilization.

The aortic cusps and sinus wall are then removed, leaving an approximately 3- to 5-mm cuff of the aortic wall in place.
The pulmonary autograft is harvested, along with an extension of the infundibular free-wall muscle, which is attached to it. This extra tissue is used for patching the ventriculoplasty incision.

After harvesting the pulmonary valve, cardioplegia is delivered, and any bleeding points from the area of previous dissection are controlled, either by diathermy or by over-sewing with a shallow suture. When the operation is finished, it is very difficult to control any potential source of bleeding from this area.
The aortic annulus is enlarged by incising the interventricular septum in the left-right commissure.

Resection of any endocardial fibroelastosis or ventricular myectomy is performed, if necessary. The goal is to remove any fibrous tissue that could obstruct the left ventricular outflow tract and limit the movement of the left ventricular cavity.
The pulmonary autograft is implanted in the left ventricular outflow tract using a running-suture technique. Natural alignment of autograft is preferable. The left semilunar cusp of pulmonary autograft should occupy the area of the previous left coronary cusp. The anterior semilunar cusp of pulmonary valve (autograft) is therefore orientated anteriorly, and the anterior lip of the retained infundibular free wall is used to patch the triangular area of incised interventricular septum.

It is important to reinforce the patching of the ventriculoplasty incision with pledgeted interrupted mattress sutures to minimize the risk of bleeding or aneurysm formation.
Reimplantation of the left coronary artery into the neoaortic root is performed first. Afterwards, the neoaortic reconstruction is completed by distal anastomosis with the ascending aorta using a continuous suture, followed by reimplantation of the right coronary artery into the neoaortic root.

The right ventricular outflow tract is reconstructed with the homograft or bovine jugular vein conduit.
An echocardiogram should be routinely performed after the patient has been weaned from bypass.
Supravalvar Aortic Stenosis

Introduction

Supravalvar aortic stenosis is a complex anomaly of the entire aortic root, with predominant stenosis at the sinotubular junction. It represents the least common form of left ventricular outflow tract obstruction and occurs in about 5–8% of patients. The underlying cause of congenital supravalvar aortic stenosis is a loss-of-function mutation of the elastine gene on chromosome 7q11.23. In patients with Williams syndrome, the elastine gene is deleted or disrupted together with a number of neighboring genes that probably are important for the other features of the syndrome, whereas in patients with familial, non-Williams supravalvar aortic stenosis, only the elastine gene is subjected to a loss-of-function translocation or point mutation. Patients with “‘sporadic’” supravalvar aortic stenosis either are members of a family carrying an elastine gene mutation with a subclinical phenotype or carry the elastine gene defect as a new mutation. The resulting decrease in elastine expression during development is associated with an increased number of elastine lamellae and excessive accumulation of collagen and smooth muscle cells in the vascular wall, which results in formation of severely thickened and rigid arterial vessels. Elastine arteriopathy is not a localized process, but rather a generalized disease of both the systemic and pulmonary arterial systems.

Anatomy

Supravalvar aortic stenosis is categorized broadly as either diffuse or localized. In the localized type, there is a severe narrowing of the sinotubular ridge. The outer diameter of the aorta may be normal or reduced, giving an hourglass appearance to the ascending aorta. The coronary arteries might be dilated either due to the high systolic pressure proximal to the sinotubular junction or stenotic due to the thickening of the wall of the sinuses of Valsalva, or they develop atherosclerosis. On occasion, the free edge of the aortic valve cusps almost completely adheres to the sinotubular junction, thus effectively isolating the coronary artery from the sinus of Valsalva and the lumen of the aorta. Abnormalities of the aortic valve are present in nearly half of the cases. Thickening of the aortic cusps and aortic insufficiency due to the high systolic pressure and bicuspid aortic valve are most commonly presented.
In the diffuse form, the entire ascending aorta and the brachiocephalic vessels might be thickened, with a narrowed lumen. There might be supravalvar narrowing of the main pulmonary artery and central and peripheral pulmonary artery stenosis.

**Indication for Surgery**

An unfavorable natural history, due to accelerated coronary artery atherosclerosis, damage of the aortic valve, and the high risk of sudden death, is a reasonable argument for proceeding with early surgery, before the left ventricular hypertrophy has become severe. A peak Doppler gradient >40 mmHg, aortic insufficiency, and compromised coronary artery blood flow are indications for surgery.

**Approach and Cardiopulmonary Bypass Strategy**

The heart is approached through a median sternotomy. The standard technique of cardiopulmonary bypass, including bicaval cannulation and mild hypothermia (32°C), are used. One has to be careful when cannulating the aorta; the aortic wall is usually thick and has a relatively small internal lumen. Placing the aortic cannula might nearly obstruct the aorta completely, with subsequent circulatory collapse and fibrillation of the heart. Myocardial protection is preferentially provided by retrograde cardioplegia; however, the first dose can be delivered directly to the coronaries. A left ventricular vent is inserted through the entrance of the right pulmonary veins.

**Symmetric Three-Patch Technique with Central Pulmonary Artery Plasty**

**The Goal of Surgery**

In the case of extensive narrowing of the left coronary sinus, a three-patch technique is preferable, providing symmetric enlargement of the aortic root. The risk of developing postoperative aortic regurgitation is minimal, despite the significant enlargement of the diameter of the sinotubular junction.
Patient Characteristics

**Age at operation:** 5 months

**Diagnosis:**
1. Supravalvar aortic stenosis
2. Severe peripheral and central pulmonary artery stenosis (hypoplasia)
3. Williams syndrome
4. Status post-balooning of the left and right pulmonary artery

**History:**
Failure to thrive with progressive congestive heart failure

**Procedure:**
1. Symmetric three patch enlargement of the aortic root
2. Pericardial patch enlargement of the central pulmonary artery

Specific Steps of Surgery

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Preoperative angiography.
Clip2
External morphology of the heart.

Clip3
Transection of the aorta and vertical incision of all three sinuses.
Three teardrop-shaped patches are sutured into each of the sinuses of Valsalva.

Patch enlargement of the central pulmonary artery.
Clip6
Reconstruction of the ascending aorta.

Clip7
Postoperative findings.
Postoperative echocardiogram.
Symmetric Nonpatch Technique (Myer’s)

The Goal of Surgery
Enlargement of the aortic root without the use of prosthetic material, preserving growth potential.

Patient Characteristics

<table>
<thead>
<tr>
<th>Age at operation:</th>
<th>9 years</th>
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<tbody>
<tr>
<td>Diagnosis:</td>
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<tr>
<td>1. Supravalvar aortic stenosis with a peak gradient &gt;50 mmHg</td>
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<tr>
<td>2. Ostial stenosis of the left coronary artery</td>
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<tr>
<td>3. Williams syndrome</td>
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<tr>
<td>History:</td>
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<tr>
<td>Syncope</td>
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<tr>
<td>Procedure:</td>
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<tr>
<td>Reconstruction of the ascending aorta using Myer’s technique</td>
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</table>

Specific Steps of Surgery

The aorta is transected at the narrowest point above the commissures. Any thickened area on the distal aorta is excised. Vertical incisions are made, similar to those in a three-patch technique, into the sinuses of Valsalva that are affected. Care is taken to avoid the coronary ostia; thus, the incisions are made off-center of the sinuses. Complementary vertical incisions are made in the distal aorta, out of phase with the proximal incisions. This allows interdigitation of the proximal and distal flaps when the aorta is reanastamosed. A zigzag anastomosis enlarges the aortic root.
Recommended Reading


