Surgical Treatment of Subaortic Stenosis:
A 17 Year Experience

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SUBAORTIC OBSTRUCTION can occur in a wide range of anatomic and functional situations and can assume many anatomic aspects, involving the left outflow tract itself and for the mitral valve as well.

This is due to the fact that embryologically in the normal heart, the left ventricular outflow tract does not develop by itself as a separate entity. But there is a developmental link between outflow and inflow parts of the heart, and we know since the studies of Lamers that this link essentially occurs at the level of the inner curvature of the heart. As pointed by Lamers, "it is the remodeling of the myocardial interventricular junction in the inner curvature that is responsible for the formation of the right atrioventricular junction and also for the formation of an outlet from the left ventricle".23)

Immediately after the completion of the loop around 25 days of intrauterine life, the heart looks like this: the outflow part is entirely located above the developing right ventricle, and the inflow part or AV canal is entirely located above the developing left ventricle. Then occur simultaneously a leftward migration of the outflow tract, and a leftward bend of the anterior part of the septum, associated with growth of the ventricles leading the AV canal rightwards. These movements result in the convergence of the outflow and inflow tract, which are above each other while the ventricular growth and septation are progressing. Together with the formation of the conal septum, by fusion of the conal ridges, between 45 and 49 days of life, occurs the wedging of the left part of the conus between mitral and tricuspid valve. This stage is necessary for achievement of a normal ventricular septation, permitting the fusion between the conal septum and the ventricular septum.

An anomaly at the stage of wedging leads to malalignment of the outflow tract relative to the ventricles. After the wedging, occurs the incorporation or absorption of the subaotic part of the outflow segment into the left ventricle, which leads the future aortic valve posteriorly and inferiorly, close to the mitral valve.

The inner curvature, or conoventricular flange, or primary fold, contributes to the formation of the left outflow tract but also of the right outflow tract. It first separates the aorta from the mitral valve then is absorbed or for Wenink, contributes to the formation of the anterior mitral leaflet. The muscle of Moulaert is considered as a remnant of the inner curvature. On the right side, it contributes to the formation of the anterior leaflet of the tricuspid valve and also to formation of the parietal band.

The formation of the outflow tract is then closely related to the development of the AV valve region. The atrial surface of the AV valves takes its origin from the AV cushions, but the ventricular surface has a myocardial origin, from the inner curvature and from the ventricles by delamination.

On the other hand intracardiac anomalies present in complex cardiac defects may lead to the absence of growth of the normal LVOT or to its development above the right ventricle. In such cases, the only outlet of the left ventricle is a VSD, as observed in double outlet right ventricle.

So we will focus during this presentation only on patients with intact ventricular septum,

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concordant ventriculo arterial connection, and then potential candidates for a biventricular repair.

Anatomy

In the normal heart, the LVOT is defined as a cone from which the base is the aortic annulus and the apex is the apex of the left ventricle. The length of the LVOT is equal to that of the inflow tract and separated from it by the anterior mitral leaflet. The anterior leaflet of the mitral valve normally extends in a straight line from the posterior to the anterior border of the left ventricle. The outflow tract of the left ventricle is bounded medially by the septal wall, and laterally by the anterior mitral leaflet and its papillary muscles. The aortic orifice, on the right, cranially and anteriorly, dominates the left ventricular outflow tract.

Recently Anderson emphasized the anatomic complexity of the aortic valve. There is no evidence to support the notion that a discrete ring supports the entire hinge point of the leaflets; on the other hand there is a discrepancy between the semi-lunar attachment of the aortic leaflets, and the ventricular aortic junction which takes the form of a circle.

In 1976 Moulaert and Oppenheimer-Dekker pointed out the fact than in about 40% of cases the anterior leaflet of the mitral valve does not reach the anterior border of the left ventricle because of the presence of a muscle bundle bordering on the left coronary semilunar cusp and extending into the anterior lateral wall of the left ventricle. The term of anterolateral muscle bundle of the left ventricle was used to name this structure which was associated with a posterolateral deviation of the mitral valve. Hypertrophy of this muscular structure can participate to a subaortic obstruction of the left ventricle.

On the other hand, this structure can participate to a complex anomaly including ventricular septal defect, biventricular origin of the pulmonary trunk and subaortic obstruction. From the right ventricle, the VSD lies immediately below the pulmonary orifice, which overrides the ventricular septum.

The outflow tract of the left ventricle is divided into an anterior and posterior chamber by a muscular ridge that arises from the anterolateral wall of the left ventricle and terminates in the ventricular septum separating a narrow aortic orifice from the large subpulmonary VSD.

Surgical Classification

Left ventricular outflow tract obstruction covers a wide range of anatomical lesions. Since the publication by Edward in 1965 several classification have been reported dealing with the anatomy of subaortic stenosis. From a surgical standpoint, subvalvar aortic stenosis can be separated into five categories, in which two main groups can be individualized: the discrete subaortic stenosis and the tunnel subaortic stenosis, with or without aortic valve anomalies.

On the other hand subaortic stenosis can be due to a mechanical obstruction related to protrusion of an anatomic structure independent from the normal LVOT.

Finally left outflow obstruction can be part of a more complex multilevel LV obstruction.

Subaortic Obstruction with Normal LVOT

Subaortic obstruction with normal LVOT includes:

- Accessory tissue of the mitral valve.
- Cardiac tumors.

Accessory mitral valve tissue causing left ventricular outflow tract obstruction is an uncommon pathologic finding in normally connected hearts. The accessory mitral tissue can form a cup shaped diverticulum on the ventricular surface of the anterior mitral leaflet bulging in systole, or an aneurysmal accessory leaflet, with a split like fenestration (Figs. 1-3).

Rarely accessory tissue of the mitral valve may be represented by masses present in the
subaortic area obstructing the pathway of the aortic valve. This pathology can be associated with a classic fibromuscular subaortic stenosis. It may remain unrecognized preoperatively, resulting in an incomplete surgical resection and a high incidence of reoperations.

Resection of the anomalous tissue is usually performed through a transaortic approach, however several groups reported an unsuccessful management using this approach, and a left atriotomy had to be performed.

Primary cardiac tumors are rare, an incidence of 0.05% was reported on the group of exclusively pediatric population. Up to 90% of primary pediatric tumors are considered as histologically benign. Among these anomalies, cardiac rhabdomyomas are the most common primary tumors in infants. These tumors are often multiple and associated with tuberous sclerosis of Bourneville in 83% of patients of the series from the Hospital of Sick Children in Toronto. Spontaneous regression of such anomalies is common, surgical resection is indicated when life-threatening symptoms, in case of increase of the tumor, or in patients with significant hemodynamic obstruction.

Amount the 16 patients treated surgically in our institution in a 16 years period 5 patients had

![Fig. 1 Echography study.](image1)
Parasternal long axis view showing an abnormal echogenic mass emerging from the anterior mitral leaflet and producing an obstruction of the LVOT in systole.

![Fig. 2 Operative view showing accessory mitral valve tissue bulging in the left outflow tract.](image2)

![Fig. 3 Same patient.](image3)
After removal of the abnormal mitral tissue, the operative picture shows abnormal subvalvar apparatus of the mitral valve.
rhabdomyoma (33% of benign tumors). Indication for surgery was in 3 cases a significant obstruction of the left outflow tract. A transaortic approach was used in all cases with good results.

Anterograde surgical technique consisting in inspection of the left ventricular cavity through the mitral valve was recommended by the Toronto group in case of mobile tumor when the tumor is inserted near the apex of the left ventricle, or located at the level of the left atrioventricular junction. More recently Ross procedure has been performed in a neonate in whom cardiac tumor involved the aortic valve.

Fibroma was the most common benign tumor in our series of patients (40%) and was present in 6 patients—3 in the right ventricle and 3 in the left ventricle. Although all the fibromas are benign, tumors of the right ventricle were resected completely and successfully. In the left ventricle localization the tumors was so huge that even partial resection was impossible without damaging the left ventricular function. In these cases as reported in the literature, cardiac transplantation may be the only alternative.

**Discrete Subaortic Stenosis**

Discrete subaortic stenosis was first reported by Chevers in 1842 and two distinct anatomical types have been identified: membranous or fibromuscular. When a fibrous ridge is firmly adherent to an hypertrophied septum anteriorly and to the left. Kelly in 1972 classified subaortic membrane in two categories: type I discrete membrane immediately under the aortic valve, and type II in which membrane is located 1 cm below the aortic valve. The shape is a fibrous ring with a stenotic orifice central and circular, or eccentric. In some cases the fibrous ridge can be thicker with the shape of a long tunnel difficult to separate from some forms of subaortic tunnel. Myocardial hypertrophy as a result of hemodynamic stenosis is more or less severe and is generally pronounced at the septal insertion of the membrane. Hypertrophy in some cases may contribute for an important part to the stenosis of the left outflow tract.

Some years ago Sommerville suggested that the so called fibrous diaphragm and discrete forms of subaortic stenosis are none of these. Actually surgical or specimen examination of the left outflow tract indicates that in most patients tissue obstructing the left ventricular outflow tract was considerably more diffuse and less membranous than anticipated. Freedom and colleagues found the typical fibrous or fibromuscular variety to be the least common in infancy.

On the other hand, as pointed out by Freedom, although cross sectional echography imaging is the best diagnostic tool for evaluation of this anomaly, it too often failed to show the true extend of the disorder so that an accurate assessment will be mandatory during surgery.

**Coexisting Cardiac Anomalies**

Discrete subaortic stenosis occurs as an isolated anomaly in about one half to two thirds of patients coming to operation. However additional anomalies are common, and have to be detected during preoperative assessment.

*Aortic valve anomalies*

The aortic valve is usually entirely normal, or with some diffuse leaflet thickening. Nevertheless trivial mild incompetence is observed in about two thirds of cases. This aortic insufficiency may be due to the anomalies of stream of blood flowing through the stenosis and damaging the cusps, but also to strands of fibroelastic tissue connecting the fibrous ridge to the aortic cusps. The aortic valve may be bicuspid and there may be congenital commissural fusion producing various degrees of valvar stenosis. On the other hand, not infrequently the valve may have been damaged by endocarditis resulting in severe incompetence, requiring an aortic valve replacement.
Mitral valve anomalies$^{15,18,19,26,35}$ (Fig. 4)

Subaortic stenosis may also be the consequence of abnormal mitral insertion accessory tissue, abnormal insertion of papillary muscle, abnormal bands within the RVOT. Abnormal septal attachment of the anterior leaflet of the mitral valve can also constitute to subaortic obstructive and are not always curable by surgery.

Muscular bundle anomalies of the LV (Fig. 5)

Posterior displacement of the infundibular septum without VSD can be a cause of subaortic obstruction associate or not to a subaortic membrane. On the other hand antero lateral muscle bundle can be an unusual cause of LVOTO.

Natural Evolution$^{37,43}$

As pointed out by Freedom the fixed forms of left ventricular outflow tract obstruction have features of both congenital and acquired disorders of the heart. Although it is generally admitted as an acquired and progressive lesion, the rate of progression is variable: particularly rapid in some patients as mild and quiescent in other cases.$^{13,14}$

Several theories have been advanced to explain the acquired nature of discrete subaortic stenosis. It is rarely present in neonates and young children and has not been reported antenatally$^{12}$. Recently, several rheological echodoppler based studies have tried to elucidate the pathological process for development of discrete subaortic stenosis.$^{2,16}$ Cape et al.$^{6}$ proposed a four-stage etiology for the development and progression of discrete subaortic stenosis. Subtle morphologic abnormalities in the left ventricular outflow tract such as a steeper aortoseptal angle result in an altered septal shear stress which triggers genetic predisposition leading to cell proliferation and structures in the LVOT. Therefore any surgical treatment should in addition to relief the subaortic stenosis attempt to treat the anatomic abnormality causing increased septal shear stress. Considering this etiological theory, it is not surprising that recurrence after resection of subaortic membrane only is a common feature in most of the published series.$^{8}$

There is a little controversy with respect to the necessity for surgical intervention. Controversies remain, however, regarding surgical timing and technique. Although early surgical removal of subaortic membranes with or without septal myotomy or myectomy seems to prevent the development of aortic insufficiency, recurrence still remains a long-term complication$^{41}$. Myectomy has been favored by several authors who demonstrated better initial and long-term follow-up$^{24}$; in our experience we are not able to find any difference between myotomy and myectomy in either the
early or the late postoperative gradient. Some authors recommended aggressive surgical approach in first intention (Hanley et al., 1999; Lecompte, 1999) but the risk of postoperative complications including complete AV block or mitral valve injury is high. On the other hand, timing of surgical repair is a matter of discussion and until now there is no consensus of the best protocol to prevent restenosis. Our opinion is similar to the policy of many groups considering that the surgical procedure is indicated when the outflow tract gradient is superior to 50 mmHg. All authors agree that residual immediate postoperative gradient, high preoperative gradient and absence of correction of associated anomalies are important factors influencing the recurrence of subaortic stenosis and aortic valve regurgitation. In all cases it appears that the relief of subaortic stenosis ultimately improves aortic valve function.

**Tunnel Subaortic Aortic Stenosis**

The long fibrous tunnel form of subaortic stenosis was described by Spencer in 1960 and was later reemphasized by Reis, Morrow and colleagues.

This anomaly presents as a circumferential irregular zone of fibrosis beginning at the end or close to the aortic valve ring and extending downwards for 10-30 mm.

Tunnel stenosis has varying degrees of severity: In the most severe forms, the stenotic tunnel is long and the diameter of the aortic annulus is small even though the aortic valve cusps are normally formed.

The left ventricular outflow tract and cavity are severely narrowed, however the ventricular septal thickness is equal to that of the left ventricular free wall.

The tubular left ventricular outflow tract narrowing remains relatively unchanged during the cardiac cycle. However some forms can be difficult to separate from asymmetric septal hypertrophy.

In less severe situations, the tunnel may be shorter and the aortic annulus normally sized resulting in difficulties to individualize these anomalies from unusual forms of discrete subaortic stenosis.

Fibrous stenosis is sufficiently long to justify the term of tunnel in about one fifty of cases of congenital subvalvar aortic stenosis and the full blown entity with annular hypoplasia is rare.

Complete form of tunnel subaortic stenosis represents a more severe and more challenging cause of left ventricular outflow tract obstruction particularly when symptoms start early in life. Early surgical intervention is generally conservative and residual stenosis is frequent, leading to reoperations. When hypoplastic aortic annulus is associated, many surgical procedure were described, besides the left ventricular apico-aortic conduit, different techniques of aortoventriculoplasty were reported.

The first and only logical approach was reported in 1975 by Konno et al. in Japan and Rastan in Germany: the surgical technique includes an aorto ventriculoplasty and an aortic valve replacement.

Size of prosthetic valve and anticoagulation required by mechanical valves are limiting factors in young pediatric population.

The basic concepts of surgical management remain the same, but the surgical procedure has evolved with time. In 1987 Clarke use an homograft instead of a mechanical valve and the anterior mitral leaflet of the homograft was used in order to augment the interventricular septum.

More recently the Ross procedure was applied to the Konno operation by Quaegebeur, Daenen Hanley and many other groups in the world. The Ross Konno procedure can be performed, even in young babies, and it is the best solution for management of these complex anomalies.

When the lesion described is a tunnel with normal aortic annulus, or in patients with recurrent subaortic stenosis, conal enlargement procedure is effective for the relief of various forms of diffuse subaortic stenosis and can be safely performed in infants and children. Vouhé et al. in 1984
reported the first series of septoplasty: he used a wide aortoseptal approach. Cooley in 1986\(^9\) reported an aortic and right ventricular approach for the conal enlargement (the modified Konno operation). Actually modified Konno operation seems to be the appropriate surgical technique.\(^10\) Preservation of the native aortic valve minimizes reoperation and long-term anticoagulation.

**Subaortic Obstruction and Complex Cardiac Anomalies**

Since the initial description of the developmental complex of parachute mitral valves, supravalvar ring of the left atrium, subaortic stenosis and coarctation by Shone and associates in 1963,\(^38\) this association has remained a therapeutic challenge for clinicians and surgeons.\(^3\) This syndrome was extended to multiple staged left ventricular obstruction. The results of medical and surgical approach have been disappointing with unsatisfactory operative outcome and poor long-term prognosis. In fact, under this terminology, there is a large spectrum of left ventricular malformation, which is classified by Kirklin, and coll. as hypoplastic left heart. In the classification of Kirklin, the severity of the malformation increases from 1 to 4 according to the number of obstructive left ventricular lesions and the extent of left ventricular hypoplasia.

Classically the approach to this complex has been to treat the symptomatic lesion delaying the repair of the other when they will become symptomatic. Therefore the initial surgical approach has been to treat the coarctation in the neonatal period or early in infancy and later the intracardiac lesion.

The real question is what to do on a sick neonate who associates several levels of left ventricular obstruction and hypoplasia of the left ventricle. We have already demonstrated that in this subset of patients the different indices that were defined for decision making between uni or biventricular repair were not relevant and the main issue was technique namely how to relieve the different levels of LV obstruction. In our series a high percentage, close to 90\%, were manageable with a biventricular repair and good outcome.\(^36\)

With regard to the subaortic obstruction in the Shone's complex, it is generally made by a membrane and the surgical decision making is similar to other type of subaortic stenosis. However, we believe it is important when embarking on an intracardiac repair to treat all levels of obstruction, namely outlet but also inlet LV obstruction.

**Patients and Methods**

From January 1980 to June 1997, among children with any form of subaortic stenosis who were referred to our Institution for surgical relief, 160 presented with intact ventricular septum, concordant ventriculo arterial connections and were candidates for biventricular repair.

There were 107 males and 53 females. The median age at intervention was 10 years (Range: 0.1-30 years). The majority of patients (126) were asymptomatic in NYHA class I–II and 34 were in NYHA class III–IV. Pre operative assessment was performed by angiography and echocardiography in 101 patients and by echocardiography alone in 59.

Gradient across the left ventricular outflow tract was calculated at catheterization by non simultaneous peak to peak systolic gradients. For patients having only echodoppler assessment the Beekman formula was applied to predict catheter-derived peak to peak LVOT gradients.

Mean gradient across the LVOT was 80±34 mmHg. Aortic valve regurgitation was present in 57 patients.

It was mild in 41, moderate in 15 and severe in 1. Aortic valve stenosis was present in 24 patients and 14 had an hypoplastic aortic annulus. Mitral valve stenosis was found in 13 patients and 9 had mitral valve regurgitation. The subaortic lesion responsible of the gradient was found to be an isolated localized discrete fibrous stenosis (membrane) in 39 patients, the latter was associated to localized muscular hypertrophy in 81 other patients. Long segment diffuse tunnel subaortic stenosis was present in 34 patients, 15 of whom having also a subaortic membrane. In 6
patients, the subaortic stenosis was particularly due to an abnormal insertion of mitral papillary muscle or to accessory anterior mitral leaflet tissue.

Surgery was indicated in presence of symptoms or an aortic regurgitation whatever the gradient across the LVOT. In asymptomatic patients decision for surgical repair was taken when the peak systolic LVOT gradient was 50 mmHg or more.

Previous surgery was performed in 33 patients including 26 coarctation repair, 2 aortic valve commissurotomy and 2 partial AV canal repairs.

**Operative Procedures**

Surgery was always conducted under hypothermic continuous cardiopulmonary bypass. Myocardial protection was ensured by antegrade and/or retrograde crystalloid or blood cardioplegia. The mean cardiopulmonary bypass time was 70.12±43.4 min and the mean cross clamp time was 32.2±20.4 min. The obstructive lesion was approached through an oblique aortotomy in all patients. In 7, a combined approach through aortotomy and right ventriculotomy was necessary.

Isolated membranectomy was performed in 39 patients, it was associated to septal myotomy in 77 and to myectomy in 26. At initial operation, it was the rule to remain as conservative as possible even leaving untouched and hypoplastic aortic annulus. Seven patients with long segment diffuse tunnel subaortic stenosis and normal aortic valve underwent through a combined approach had a modified Konno procedure by patch septoplasty as the initial operation.

Three patients with tunnel subaortic stenosis and hypoplastic aortic annulus had a Konno-Rastan procedure and two other with the Shone’s syndrome had apical conduit insertion. In six patients, the treatment of subaortic stenosis consisted in resection of an accessory mitral valve tissue. Aortic commissurotomy was associated in 15 patients and 6 others had aortic valve replacement.

Mitral valve repair was performed in 4 patients and two other underwent mitral valve replacement.

**Follow-up**

All but 10 foreigner patients were regularly reviewed by their referring cardiologist achieving a median follow-up 13.3 years (Ranges: 1.2-17.9 years). Particular attention was given to the recurrence of subaortic stenosis and to the function of the aortic and mitral valves.

**Results**

**Mortality/Morbidity**

There were 5 early (3.1%; 70%CL: 1.5-5.2%) deaths. Among them, 3 patients presented with tunnel subaortic stenosis, 1 with already compromised left ventricular function (NYHA III-IV) and the two other had severe preoperative aortic valve regurgitation. One of the latter underwent unsuccessfully a Konno-Rastan aortoventriculoplasty with mechanical aortic valve replacement. Two other post operative deaths occurred in patients with discrete subaortic stenosis, one from a iatrogenic severe aortic regurgitation and the other from secondary tamponade. At univariate statistical analysis, only pre operative NYHA functional class III-IV and the older age at operation significantly increased the risk for early death (p=0.0002 and 0.006, respectively), however none of these variables were independent risk factor at multivariate analysis. Five patients required pace maker implantation for complete AV block as a result of surgery.

**Aortic Valve Function**

Seventy six patients were known preoperatively to have aortic valve dysfunction. Nineteen had isolated aortic valve stenosis, 51 had isolated aortic valve regurgitation (AR) and 6 had aortic valve disease with both regurgitation and stenosis. The degree of aortic regurgitation at presenta-
tion correlated with age ($p=0.04$) but not with LVOT gradient. Relief of the subaortic stenosis improved AR degree in 49 patients, stabilized it in 4 patients but it was worsened in 2 with preoperative AR and appeared in 21 without preoperative AR. However, in the latter, AR was mild and only one was reoperated on for aortic endocarditis. All the other did not demonstrate aggravation of the AR during the length of follow-up. No preoperative factors were found to influence the outcome of aortic valve function (age, preop. gradient, surgical technique, anatomy, preoperative valve function).

**Post Operative Mitral Valve Function**

Preoperative mitral valve stenosis was diagnosed in 13 patients. It was mainly due to papillary muscle fusion, however in only 3, it was necessary to associate a procedure for relief of mitral stenosis. Mitral regurgitation was present in 9 patients before surgery, 2 underwent mechanical mitral valve replacement and 1 had mitral valvuloplasty. Eleven patients developed a mild to moderate mitral regurgitation following surgery.

**Recurrence of Subaortic Stenosis and Reoperations**

The gradient across the LVOT before initial operation did not correlate with preoperative age ($r=0.11$, $p=0.08$). It was reduced in all survivors from $80 \pm 34$ mmHg to $16 \pm 3.5$ mmHg. No significant difference in reduction of the LVOT gradient was observed relative to preoperative anatomy, LVOT gradient, to association with aortic valve disease to surgical technique and to age. Particulary, there was no difference when myotomy was performed versus myectomy.

All traced patients had an available echodoppler study within 1 year of the completion of this study. Forty two (27.1%; 70% CL: 23.2-31.3%) patients developed a gradient superior to 30 mmHg across the LVOT. The mean delay for reappearance of a gradient across the LVOT was 3.65 ± 3.35 years. Statistical analysis revealed that several factors could be predictors for recurrence of the subaortic stenosis. At univariate analysis, anatomical, surgical and hemodynamic factors were strong predictors for recurrence. They were: relative hypoplasia of aortic anulus ($p=0.02$), tunnel form ($p=0.018$), existence of aortic coarctation ($p=0.0008$); myectomy and isolated membraoctomy were associated with a higher rate of recurrence ($p=0.03$ and 0.02, respectively) and higher pre gradients across the LVOT ($p=0.05$). There was a strong positive correlation ($r=0.3$, $p=0.0001$) between immediate post operative LV to aorta gradient and the occurrence of recurrence. At multivariate Cox regression analysis, existence of aortic coarctation ($p=0.005$) and the immediate post operative gradient across the LVOT ($p=0.005$) were independent risk factors for recurrence.

Twenty patients (12.9%; 70% CL: 9.96-16.3%) were reoperated on within a mean delay of 5.35±4.6 years. Recurrent subaortic stenosis was the cause for reoperation in 17 (10.9%; 70% CL: 8.2-14.1%) with a mean gradient across the LVOT of $95 \pm 29$ mmHg. Fourteen underwent a new membrane excision associated to myectomy, 1 had a mechanical aortic valve replacement, 2 had a Ross-Konno procedure, 1 had an apical conduit insertion and 2 underwent apical conduit replacement. There were no early death after reoperation. Six of the previous patients underwent a second reoperation for aortic prosthetic valve endocarditis in 1, recurrent subaortic stenosis in 4 and apical conduit obstruction in 1.

Statistical analysis revealed that reoperation rate was influenced at univariate analysis by the presence of an hypoplastic aortic anulus ($p=0.014$), presence of an aortic coarctation ($p=0.005$), younger age at initial operation ($p=0.014$) (Fig.6) and the immediate post operative gradient across LVOT ($p=0.0001$).

**Overall Mortality and Long Term Follow-up**

There were four late deaths (4.37%; 70% CL: 2.57-6.75%), two occurred after a second reoperation, one who underwent apical conduit insertion at reoperation died 6 months later from
severe myocardial dysfunction and the latter died from organic chronic renal failure. Overall mortality was essentially influenced by anatomical factors namely: hypoplastic aortic anulus \( (p=0.0001) \), mitral stenosis \( (p=0.0026) \), tunnel form of subaortic stenosis \( (p=0.01) \) and existence of coarctation \( (p=0.018) \). Multivariate analysis showed that hypoplastic aortic anulus \( (p=0.01) \) and mitral stenosis \( (p=0.048) \) were independent predictors for overall mortality.

At a median follow-up of 13.3 years, all but two survivors were in NYHA class I–II and two were in NYHA class III–IV. Two other patients have successfully underwent liver transplant. The mean gradient across the LVOT was 20±13 mmHg. Twenty two patients had an aortic valve insufficiency, it was mild in 11 and moderate in 11.

Actuarial survival at 15 post operative years was 94.25±1.34% and freedom from reoperation was 85.3±6% (Figs. 6–7).

In conclusion, subaortic stenosis covers a large range of anatomic anomalies requiring a careful preoperative and intraoperative assessment. Echodoppler has to take into account the main lesions and also the additional anomalies which must be treated during operation. Discrete subaortic stenosis can be cured in the majority of patients by membranectomy associated to either myotomy or myectomy. However, because the anatomic substrate is not addressed by these surgical techniques, recurrences are likely to occur during the long term follow-up, particularly in patients who has been previously operated on for an aortic coarctation and those who had a less than optimal relief of LVOT gradient. In this subset of patients, the optimal surgical technique remains to be described but requires probably a more aggressive approach. Tunnel subaortic stenosis represents a more severe and more challenging cause of left ventricular outflow tract obstruction. According to the size and function of the aortic valve, the Ross-Konno procedure or the modified Konno by patch septoplasty seems to be the appropriate surgical techniques. In all cases, it seems that relief of the subaortic stenosis improves the fate of aortic valve function.

References

3) Black, M. D., Kadletz, M., Smallhorn, J. F. et al.: Cardiac rhabdomyomas and obstructive left heart


