

# State of the art of cardiac surgery in patients with congenital heart disease

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During the last 20 years, pediatric cardiac surgery has been characterized by important changes, with reductions in surgical mortality and the achievement of complete repair at an earlier age, thus avoiding multiple procedures and strongly ameliorating the global outcome of these patients. In this review, we describe the actual trends in the surgical treatment of cardiac malformations. We analyze two groups of patients: in the first group (septal defects, tetralogy of Fallot, transposition of the great arteries, aortic stenosis and coarctation) the indications are well established and the goal is represented by a lessening of the surgical trauma and post-operative morbidity, with stable results in the follow-up. In the second group (univentricular heart, pulmonary atresia and intact ventricular septum, double

discordance, conduit, hypoplastic left heart syndrome), the lesions are still considered complex and submitted to ongoing experimental and clinical research, in order to improve the post-surgical history of these diseases. *J Cardiovasc Med* 8:3–6 © 2007 Italian Federation of Cardiology.

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## Introduction

The movement of cardiac surgery to the very young was the title of the keynote address of Dr Kirklin at the first World Congress of Pediatric Cardiac Surgery in Bergamo in 1988. Pediatric cardiac surgery, in the last 20 years, has been characterized by the treatment of common heart malformations in the neonatal period, thus avoiding multiple procedures and strongly ameliorating the global outcome of these patients. The surgical treatment of the most common forms of congenital heart disease is nowadays sufficiently standardized to obtain good results with low mortality and morbidity and favorable hemodynamic outcomes in the long term. In the first part of this review, we will analyze the group of congenital heart diseases in which the goal is no longer represented by a reduction of mortality, but by a lessening of the surgical trauma and post-operative morbidity, with stable results in the follow-up.

## Septal defects

In such defects, both at the atrial and ventricular level, interventional cardiology is now widely and successfully applied. Inter-atrial defects are still 'surgical' when they are too large or lack borders for safe device positioning. Ventricular septal defects (VSDs) are of surgical interest if they need closure in the first 6–12 months of life. Therefore, in this field, the evolution of the surgical approach is represented first by attempts to reduce the surgical trauma and the post-operative discomfort, shorten hospital stays with reduced utilization of hospital resources and ameliorate the aesthetic results

(mini-invasive techniques). Furthermore, regarding the treatment of multiple and complex muscular VSDs, the new and emerging combined approach between surgery and intra-operative device application, should allow a less aggressive surgery with reduced myocardial ischemia and the opportunity to perform correction in smaller infants with better and more stable results [1,2]. Regarding the atrioventricular (AV) defects, closure of the 'cleft' in the left AV valve is now widely applied and accepted. Surgery should be performed during the first 6 months of life in patients with a complete AV canal. In partial AV defects, repair in the first 12–24 months of life is strongly recommended to avoid acquired stress lesions of the valve leaflets and progressive annular dilatation, leading to worst surgical outcomes. The mortality is now generally under 5%, but an incidence of 10–20% of re-do surgery [3] is still reported for left AV valve residual problems (mainly regurgitation).

## Tetralogy of Fallot

Correction during the first 6 months of life is recommended and can be achieved with a low mortality (< 5%). Some controversies still remain regarding primary neonatal repair, irrespective of age and body weight, versus early palliation. A still unsolved problem is to quantify the need for a complete relief of the right-ventricle-to-pulmonary-artery gradient at the time of repair [4], leaving a small and well tolerated residual infundibular or valvular obstruction, thus allowing for a small size compared with the standard strategy. The results are satisfactory, and there has been speculation on the potentially better

long-term results due to the absence of pulmonary valve regurgitation, right ventricular dilatation, and the need for future pulmonary valve replacement. Other authors [5,6] reviewed the long-term results after the repair of tetralogy showing acceptable results but with a substantial incidence of right ventricular dilatation and ventricular arrhythmias secondary to pulmonary valve regurgitation, which sometimes need re-operation.

### **Transposition of the great arteries**

The surgical treatment of transposition of the great arteries (TGA) in the 'switch' offers good early and long-term results, with a low (< 5%) operative mortality in the simple forms; however, complex forms (like TGA with VSD and coarctation) still remain challenging. In this group, clinical research has been applied to the field of extracorporeal circulation technology, aiming to reduce the effects of the capillary leak syndrome and cerebral damage. Thanks to these advances, cardiopulmonary bypass (CPB) technology can now be used for the correction of many other complex forms of congenital heart disease in the neonatal period.

### **Aortic stenosis**

The treatment of valvular aortic stenosis with percutaneous balloon dilatation had a great impact on clinical results in newborns, but treatment after the first year of age is still debated. Surgery offers the advantage of a more accurate evaluation of anatomic lesions and the possibility of more refined plastic techniques, specifically adapted for each single type of lesion. Due to the high probability of repeated procedures during life the interventional approach should be considered even beyond the first year of life. In the complex forms, especially when associated with stenosis/hypoplasia of the subaortic region or regurgitation of the aortic valve, the Ross and Ross-Konno operations have been successfully applied in neonatal and pediatric patients. In this age group, mortality is about 10% but the long-term results are much better in respect of valve replacement, mostly due to the potential for growth of the new aortic route; however, concerns still remain about dilatation of the pulmonary autograft during the immediate post-operative period, followed by normal active growth [7,8]. Treatment of subaortic stenosis is well established. Early repair is suggested to avoid acquired lesions of the aortic valve and severe myocardial hypertrophy. Diaphragm ablation by blunt dissection is always associated with myotomy/myectomy. As a marker for increased risk of recurrence, echocardiographic detection of associated anomalies of the left ventricular outflow tract, such as anomalous chordae, mitral anomalies, or muscular septal bulgings, has been proposed [9,10]. Only aggressive and early treatment can avoid possible coronary lesions and the severe left ventricular hypertrophy in supra-valvar aortic stenosis. Several surgical techniques [11] have been developed: single patch enlargement (with extension into

the non-coronary sinus) and inverted Y patch (with extension into two sinuses) have been the most utilized. Nonetheless, the frequent association with Williams-Van Beuren syndrome, with its widespread involvement of aortic wall, aortic and renal vessels, makes it almost impossible to control the evolution of the associated hypertensive disease. Aortic coarctation repair, despite the use of different surgical techniques, still presents suboptimal results when a long-term follow-up is obtained and an exercise test is performed in these patients. Hypoplasia of the aortic arch, once not addressed during surgery, is today more frequently approached in the newborn, because residual hypoplasia may be responsible for late hypertension even in the absence of residual stenosis. The shape of the aortic arch (roman or gothic arch) has been described to be correlated with late systemic hypertension and an abnormal response to stress test [12].

### **Univentricular heart**

Typically, the group of cardiac anomalies defined as 'univentricular heart' represent a difficult and challenging form of congenital heart disease. Surgical history is represented by more operations, with uncertain results in the medium and long-term follow-up, especially regarding timing for bidirectional cavopulmonary anastomosis (BCPA) and (Fontan) and the type of operation (additional pulmonary flow source and fenestration in Fontan patients). Regarding the technique of extracardiac total cavopulmonary connections, in order to provide a more efficient circulation, it is important to reduce the energy loss at the level of the connection between the caval veins and the pulmonary arteries. In-vitro studies showed that optimal flow, with minimal energy loss, can be obtained by flaring at the site of both anastomoses (superior and inferior vena cava) combined with caval offset [13]. Finally, it is important to focus on performing a BCPA or a total cavopulmonary connection without the use of cardiopulmonary bypass [14], resulting in better post-operative hemodynamics, lung function and decreased utilization of blood and its derivatives.

### **Pulmonary atresia, intact ventricular septum**

In pulmonary atresia with intact ventricular septum, the surgical choices, even today, are not well standardized and the long-term follow-up is uncertain [15]. The surgical treatment varies depending on the anatomy of the tricuspid valve (Z value), the right ventricle and its outflow tract, and by institutional policy. Percutaneous radio-frequency perforation and balloon dilatation of the valve are often successful in cases with adequate right ventricle and infundibulum. The sooner the right ventricular decompression is obtained, the better are the results in terms of growth of the right ventricle and the tricuspid valve. Often a temporary alternative pulmonary blood flow source is needed: a patent ductus with prostaglandin infusion or an aortopulmonary shunt. When the right

ventricle is only partially adequate, a 'one and a half ventricle' strategy may be considered, in the form of a complete separation of the pulmonary and systemic circulations, but with partial right unloading through a bidirectional Glenn procedure. This approach is now applied to an expanding number of congenital heart defects, with morphologic or functional characteristics that are not amenable to biventricular repair [16].

### Double discordance

An extensive retrospective study [17] of children with double discordance (corrected TGA) treated by the conventional approach, with a follow-up of 20 years, showed a cumulative high mortality with a substantial incidence of AV systemic (tricuspid) valve replacement, complete AV block necessitating pacemaker implantation, and the need for re-operation. This study concluded by suggesting consideration for alternative surgical management. The double switch procedure (atrial and arterial), which was reported for the first time in 1990, has been applied with an expanded indication worldwide, with acceptable early and mid-term results. Although appealing, it is a technically demanding and long surgical procedure and still has a significant surgical mortality; furthermore, the long-term outcome (and particularly the atrial switch component of the operation) is not yet clarified. Follow-up studies are needed to compare the long-term results of conventional surgical treatment with the double switch operation.

### Conduits

Immediate results of right ventricle to pulmonary artery conduits are usually satisfactory but, particularly in infants, almost always commit the patient to multiple surgical re-operations, due to progressive conduit obstruction. Percutaneous dilatation and stenting are successful in delaying the need for re-operation and conduit replacement [18], but usually leave a pressure gradient with concomitant pulmonary valve insufficiency; the long-term effects on the right ventricular function should be taken into consideration. Re-intervention should be planned not only on the basis of the severity of obstruction, but also depending on the right ventricular function, especially if pulmonary regurgitation is present, but the optimal timing and type of surgical replacement of the conduit is still a matter of controversy [19].

### Hypoplastic left heart syndrome

During the 1990s and up to the beginning of this century, the treatment of hypoplastic left heart syndrome (HLHS) was the landmark of surgical excellence, just as TGA had been during the previous 10 years; a demanding heart disease which has a rapidly fatal outcome without treatment. Similar to the association of TGA with the development of neonatal CPB, HLHS has endorsed the application of new techniques of cerebral protection, such as selective cerebral perfusion [20]. Recently [21],

the first stage has been performed with a beating heart, thereby avoiding circulatory arrest and myocardial ischemia. Considering the importance of myocardial preservation for the long-term results, this approach may improve the results of the management of this complex defect. The increasing experience with the Norwood procedure also helped the understanding of the delicate balance between systemic and pulmonary blood flow in the post-operative period. Furthermore, HLHS has spurred advances in echocardiographic diagnosis, including in-utero diagnosis and intervention and is the paradigm for early post-natal resuscitation of the acidotic neonate with a closing ductus. HLHS treatment paved the way toward widespread application of intermediate procedures as well as the introduction of neonatal heart transplant programs, giving rise to ethical and moral issues such as the use of anencephalic babies as heart donors and the concentration of expensive resources for the benefit of a relative small number of babies. Surgical techniques for HLHS are still evolving but no real consensus has been yet achieved, even for the decision 'to treat or to do nothing' for these patients.

### Conclusions

Although in the great majority of patients with congenital heart defects, the surgical indications, approach and techniques are today well established with good immediate and long-term results, there is still plenty of scope for cardiologic, surgical and intensive care discussion, to improve the results and the quality of care for the most complex cardiac malformations. As there is a very small frequency of these malformations (about 0.8% born but only 0.3–0.4% of surgical interest), it is mandatory to have a small number of institutions performing the cardiologic and cardiosurgical treatment for these complex patients, to improve the performance and to obtain the experience necessary to reduce mortality, morbidity and to perform statistically significant clinical studies.

### References

- 1 Bacha E, Cao Q, Stair JP, Waight D, Ebeid ME, Hijazi ZM. Periventricular device closure of muscular ventricular septal defects on the beating heart: technique and results. *J Thorac Cardiovasc Surg* 2003; **126**:1718–1723.
- 2 Amin Z, Danford DA, Lof J, Duncan KF, Froemming S. Intraoperative device closure of perimembranous ventricular septal defect without cardiopulmonary bypass: preliminary results with the periventricular technique. *J Thorac Cardiovasc Surg* 2004; **127**:234–241.
- 3 Michielon G, Stellin G, Rizzoli G, Casarotto DC. Repair of complete common atrioventricular canal defects in patients younger than four months of age. *Circulation* 1997; **96 (suppl II)**:316–322.
- 4 Rao V, Kadletz M, Hornberger LK, Freedom RM, Black MD. Preservation of the pulmonary valve complex in tetralogy of Fallot: how small is too small? *Ann Thorac Surg* 2000; **69**:176–180.
- 5 Norgaard MA, Lauridsen P, Helvind M, Petterson G. Twenty-to-thirty-seven year follow-up after repair for tetralogy of Fallot. *Eur J Cardiothorac Surg* 1999; **16**:125–130.
- 6 Oechslin EN, Harrison DA, Harris L, Downar E, Webb GD, Siu SS, Williams WG. Reoperation in adults with repair of tetralogy of Fallot: indications and outcomes. *J Thorac Cardiovasc Surg* 1999; **118**:245–251.
- 7 Luciani GB, Baroni L, Tomezzoli A, Casali G, Mazzucco A. Bicuspid aortic valve disease and pulmonary autograft root dilatation after the Ross procedure: a clinicopathologic study. *J Thorac Cardiovasc Surg* 2001; **122**:74–79.

- 8 Solymar L, Sudow G, Holmgren D. Increase in size of the pulmonary autograft after the Ross operation in children: growth or dilatation? *J Thorac Cardiovasc Surg* 2000; **119**:4–9.
- 9 Cohen L, Bennani R, Hulin S, Malergue MC, Yemets I, Kalangos A, *et al*. Mitral valvar anomalies and discrete subaortic stenosis. *Cardiol Young* 2002; **12**:138–146.
- 10 Marasini M, Zannini L, Ussia GP, Pinto R, Moretti R, Lerzo F, Pongiglione G. Discrete subaortic stenosis: incidence, morphology and surgical impact of associated subaortic anomalies. *Ann Thorac Surg* 2003; **75**:1763–1768.
- 11 Stamm C, Kreutzer C, Zurakowski D, Nollert G, Friehs I, Mayer JE, *et al*. Forty-one years of surgical experience with congenital supra-avalvular aortic stenosis. *J Thorac Cardiovasc Surg* 1999; **118**:874–885.
- 12 Ou P, Bonnet D, Auriacombe L, Pedroni E, Balleux F, Sidi D, Mousseaux E. Late systemic hypertension and aortic arch geometry after successful repair of coarctation of the aorta. *Eur Heart J* 2004; **25**:1853–1859.
- 13 Amodeo A, Grigioni M, Oppidio G, Daniele C, D'Avenio G, Pedrizzetti G, *et al*. The beneficial vortex and best spatial arrangement in total extracardiac cavopulmonary connection. *J Thorac Cardiovasc Surg* 2002; **124**:471–478.
- 14 McElhinney DB, Petrossian E, Reddy VM, Hanley FL. Extracardiac conduit Fontan procedure without cardiopulmonary bypass. *Ann Thorac Surg* 1998; **66**:1826–1828.
- 15 Jahangiri M, Zurakowski D, Bichell D, Mayer JE, del Nido PJ, Jonas RA. Improved results with selective management in pulmonary atresia with intact ventricular septum. *J Thorac Cardiovasc Surg* 1999; **118**:1046–1055.
- 16 Mavroudis C, Backer CL, Kohr LM, Deal BJ, Stinios J, Muster AJ, Wax DF. Bidirectional Glenn shunt in association with congenital heart repairs: the  $1\frac{1}{2}$  ventricular repair. *Ann Thorac Surg* 1999; **68**:976–982.
- 17 Yeh T, Connelly MS, Coles JG, Webb GD, McLaughlin PR, Freedom RM, *et al*. Atrioventricular discordance: results of repair in 127 patients. *J Thorac Cardiovasc Surg* 1999; **117**:1190–1203.
- 18 Ovaert C, Caldarone CA, McCrindle BW, Nykanen D, Freedom RM, Coles JG, *et al*. Endovascular stent implantation for the management of postoperative right ventricular outflow tract obstruction: clinical efficacy. *J Thorac Cardiovasc Surg* 1999; **118**:886–893.
- 19 Dearani JA, Danielson GK, Puga FJ, Schaff HV, Warnes CW, Driscoll DJ, *et al*. Late follow-up of 1095 patients undergoing operation for complex congenital heart disease utilizing pulmonary ventricle to pulmonary artery conduit. *Ann Thorac Surg* 2003; **75**:399–411.
- 20 Tchervenkov CI, Al-Khaldi A, Shum-Tim D. Antegrade regional cerebral perfusion. *Cardiol Young* 2004; **14** (suppl 1):70–74.
- 21 Kishimoto H, Kawahira Y, Kawata H, Miura T, Iwai S, Mori T. The modified Norwood palliation on a beating heart. *J Thorac Cardiovasc Surg* 1999; **118**:1130–1132.