

# Long-term natural history and follow-up of children operated for congenital heart disease

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Over the past 20 to 30 years, major advances have been made in the diagnosis and treatment of congenital heart disease in children. As a result, patients now survive to adulthood. Nowadays in Italy there are between 50 000 and 60 000 patients aged 1 to 21 years old and affected by congenital heart disease, 40% of whom underwent surgical repair or palliation of their defects. We will discuss the more common acyanotic and cyanotic congenital heart conditions, treated or not surgically, that physicians who care for adults are likely to encounter.

## Acyanotic conditions

**Atrial septal defect.** Patients who underwent surgical closure for isolated atrial septal defect in pediatric and juvenile age have a normal life expectancy, while closure after 40 years of age, even if improving prognosis and functional class does not reduce the risk of atrial fibrillation, present in about 30% of patients. Up to 40% of patients who did not undergo defect closure develop atrial fibrillation during follow-up<sup>1</sup>. The surgical outcome of ostium primum atrial septal defect can be different, because of the involvement of atrioventricular valves: even after the complete repair of the defect, significant mitral valve anomalies may persist, usually in the form of mild regurgitation (leading to the possible onset of atrial fibrillation in about 33% of patients during follow-up early after surgical closure). This condition may lead, in up to 10% of these patients, to the need for reoperation within

the following 10 years<sup>2</sup>. Prophylaxis against infective endocarditis is recommended for patients with a residual valvular abnormality.

**Ventricular septal defect.** Adult patients with ventricular septal defect usually have a little shunt because of the spontaneous closure or size reduction of the defect, while patients with wide defects often develop Eisenmenger's syndrome. Adult patients with a small ventricular septal defect may become symptomatic for progressive subpulmonary stenosis or insufficiency of the aortic valve due to leaflet prolapse, both needing surgical intervention<sup>3</sup>. Long-term outcome after surgery for ventricular septal defect is good. However, some patients present with residual anomalies (persistent pulmonary hypertension, ventricular arrhythmias, ventricular conduction delays, limited functional capacity at cardiopulmonary exercise testing). The age at operation is paramount to ensure a good outcome, above all for those patients with hemodynamically important defects. Patients operated under 2 years of age have no symptoms, a normal growth and a good prognosis, while those operated during adult life may develop a residual cardiac damage (reduced left ventricular function, increased left ventricular volume, high pulmonary vascular resistances)<sup>2</sup>. Significant complications are usually restricted to the short-term follow-up period, suggesting a good long-term outcome<sup>4</sup>. Patients with a residual shunt should receive prophylaxis for infective endocarditis.

**Pulmonary stenosis.** Long-term follow-up data of surgical pulmonary valvulotomy carried out in infancy are excellent in patients with elastic leaflets, en-dome opening and normal valvular ring, with a late mortality rate similar to the general population<sup>2</sup>. However patients submitted to transvalvular patch application often develop pulmonary insufficiency. In recent years, percutaneous balloon valvuloplasty has become the procedure of choice with excellent results.

**Aortic stenosis.** Patients with mild aortic stenosis who are asymptomatic have a normal life but should receive antibiotic prophylaxis against infective endocarditis. Once symptoms appear, survival is limited: therefore symptomatic aortic stenosis should undergo surgical treatment or transcatheter valvuloplasty. Surgical therapy of valvular aortic stenosis in infancy does not warrant complete recovery from the disease. Neonates with critical valvular aortic stenosis who survive surgical or transcatheter valvulotomy, often develop a recurrence of aortic stenosis or a significant insufficiency within 10 years of age. Surgical options in this group of patients are either surgical valve replacement or to consider a Ross procedure for children under 18 years of age. Surgical timing depends on hemodynamic gradient, valvular morphology, calcification or the onset of endocarditis. Nowadays percutaneous balloon valvuloplasty represents the method of choice in infancy.

**Aortic coarctation.** Surgical repair should be considered for patients (even adults) with a transcoarctation pressure gradient of > 30 mmHg. Although balloon dilation is a therapeutic alternative, the procedure is associated with a higher incidence of recurrent coarctation with respect to surgical repair in the neonatal period<sup>5</sup>. Postoperative complications include recurrent coarctation and the development of aortic aneurysm. The incidence of persistent or recurrent hypertension, as well as survival rate, is influenced by the patient's age at the time of surgery. If repair of coarctation is performed between 20 and 40 years, the 25-year survival is 75%. When repair is performed after 40 years of age, the 15-year survival is only 50%<sup>2</sup>.

**Congenitally corrected transposition of the great arteries.** Patients with congenitally corrected transposition of the great arteries without associated defects usually remain asymptomatic until 40 years. Complications observed in these patients are atrioventricular block, systemic valve (tricuspid) regurgitation (needing replacement), supraventricular arrhythmias and heart failure. Patients with associated defects (ventricular septal defect, pulmonary stenosis) need surgical treatment before adulthood<sup>3</sup>.

## Cyanotic conditions

Patients with cyanotic congenital heart disease have systemic oxygen desaturation resulting from left-to-right shunt. The magnitude of shunting determines the severity of desaturation. Most children with cyanotic heart disease do not survive to adulthood without surgical intervention. In adults, the most common causes of cyanotic congenital heart disease are tetralogy of Fallot and Eisenmenger's syndrome.

**Tetralogy of Fallot.** In the past, infants were treated by increasing the pulmonary blood flow through a systemic-to-pulmonary shunt, thereby reducing the severity of cyanosis and improving exercise tolerance. Often, however, these procedures were associated with long-term complications. Currently, complete surgical correction is performed at very young age. Mortality associated with surgery is less than 3.0% in children and 2.5 to 8.5% in adults<sup>6</sup>. Patients with tetralogy of Fallot (either repaired or unrepaired) should therefore receive prophylaxis for endocarditis with antibiotics before dental or elective surgical procedures. Although they are usually asymptomatic, survival is somewhat poorer with respect to an age-matched control population, because of an increased risk of sudden death. In one series, the rate of survival 32 years after surgery was 86% among patients with repaired tetralogy and 96% in an age-matched control population<sup>7</sup>. Patients with repaired tetralogy of Fallot often have ventricular arrhythmias or atrial fibrillation or flutter, which may cause considerable morbidity. Pulmonary regurgitation, residual or recurrent obstruction of the right ventricular outflow tract or residual ventricular septal defect require repeated surgery.

**Congenital cardiac anomalies corrected with Fontan operation.** The Fontan-like surgical repair is considered as the definitive palliation for patients with only one functional ventricle, like tricuspid atresia, univentricular heart or any other type of hypoplasia of the left or right ventricle. Medium to long-term results depend on the rigorous preoperative selection of patients standing for this kind of intervention, while short-term results are usually good. Often, patients who received a Fontan operation with an *ad hoc* selection (low end-diastolic ventricular pressure, low pulmonary artery pressure and absence of systemic atrioventricular valve insufficiency) are still in NYHA functional class I-II 10-15 years after the intervention<sup>8</sup>. In those patients, the maintenance of sinus rhythm is mandatory for a good hemodynamic balance. Supraventricular tachyarrhythmias can in fact reduce ventricular function and produce an increase in pulmonary vein pressure. In patients presenting with severe right atrial dilation and supraventricular arrhythmias in the setting of a classical atrio-pulmonary connection should be considered for a conversion to total cavopulmonary bypass. Arrhythmias

and protein-losing enteropathy are possible complications observed during follow-up<sup>7</sup>.

**Transposition of the great arteries.** Two surgical procedures have been used in patients with complete transposition of the great arteries. The initial approach, known as the atrial switch operation, is featured by an intra-atrial rerouting of the pulmonary and systemic venous flows (Mustard or Senning approach). The complications of this kind of operation are due to the leakage and obstruction of the atrial baffle, sinus node dysfunction and atrial arrhythmias (particularly atrial flutter), right (systemic) ventricular dysfunction and an increased risk of sudden death<sup>8</sup>. The 10- and 20-year survival rate is 90 and 80%, with most patients in NYHA functional class I-II<sup>7</sup>. The atrial switch operation has recently been replaced by the arterial switch operation, in which the pulmonary artery and ascending aorta are transected above the semilunar valves and coronary arteries and then switched. The coronary arteries are re-located to the neo-aorta to restore normal coronary circulation. This operation can be performed in neonates and is associated with a low operative mortality (usually < 5%) and an excellent long-term outcome<sup>7</sup>. Most complications observed are the development of constriction of the pulmonary outflow tract, aortic valve insufficiency or stenosis of coronary arteries.

**Eisenmenger s syndrome.** Eisenmenger s syndrome is due to the presence of a large left-to-right shunt that causes severe pulmonary vascular disease and pulmonary hypertension, with resultant reversal of the direction of shunting and appearance of cyanosis. Patients with congenital heart diseases develop Eisenmenger s syndrome for late surgical intervention, incomplete correction or as a consequence of palliative operations. Most patients have impaired exercise tolerance and exertional dyspnea, but these symptoms may be well compensated for years. Patients refer palpitations, often due to atrial fibrillation or flutter. They develop erythrocytosis due to arterial desaturation and symptoms of hyperviscosity and abnormal hemostasis may appear. Hemoptysis may occur, as a result of pulmonary infarction

or rupture of dilated pulmonary or aortico-pulmonary collateral vessels. Cerebrovascular accidents may occur as a result of paradoxical embolization. Patients may have syncope due to inadequate cardiac output or arrhythmia. Symptoms of heart failure, which are uncommon until the disease is far advanced, portend a poor prognosis. The survival rate among patients with Eisenmenger s syndrome is 77% at 15 years, and 42% at 25 years<sup>7</sup>. Death is usually sudden, presumably caused by arrhythmias<sup>8</sup>.

Special aspects that may involve both the general physician and the cardiologist caring patients with congenital heart disease are:

- the relation between congenital heart disease and an increased risk during pregnancy;
- hematologic problems in cyanotic cardiac anomalies, such as polycythemia and platelet dysfunction;
- indications to physical activity and sport in patients affected by congenital heart disease.

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