Simple D-transposition of great arteries operated at the age of 11 years

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The simple transposition of the great arteries is a lethal congenital heart disease. The life expectancy of unoperated patients is about 9 months.

We report the original observation of a girl with unoperated simple transposition of the great arteries, who survived until the age of 11 years. An atrial switch was successfully performed according to the technique of Senning–Mustard.

Keywords: Transposition of the great arteries, Senning, Mustard

Introduction

The transposition of the great arteries is the most common congenital heart disease (CHD) diagnosed in the neonatal period and probably the most serious. Thus, it constitutes a diagnostic and therapeutic emergency.

Case report

F.S. is an 11 year old girl, living in a rural area. She came out from a term pregnancy with a non-medical birth.

The story of her illness began at birth by the appearance of a cyanosis, more marked in the lips and extremities. This cyanosis was neglected by the family, seeing a normal growth in size and weight and a good psychomotor development of their daughter. It’s only at the age of 11, that a schoolteacher noticed her abnormal “blue” skin, and addressed her to a cardiologist.

Clinical examination found the patient in relatively good condition with normal weight and size. There was a generalized cyanosis with SaO2 at 60% and digital clubbing. Blood pressure is 100/60 mmHg and cardiac auscultation finds a systolic murmur grade 2/6 along the left sternal border.

The electrocardiogram recorded a regular sinus rhythm at 100 bpm, a right axis, right atrial and ventricular hypertrophy.

Transthoracic echocardiography revealed a D-transposition of great arteries with a dilated pulmonary artery. The two ventricles are balanced with a preserved left ventricle systolic function (LVEF = 70%). The atrial septum is multi perforated with a bidirectional inter-atrial shunt. The ventricular septum is intact and there is no persistent ductus arteriosus.
The mean pulmonary arterial pressure (PAPm) is around 15–20 mmHg. Cardiac catheterization confirmed the low pulmonary arterial pressures (PAPs = 35 mmHg; PAPd = 10 mmHg; PAPm = 16 mmHg).

Laboratory tests revealed a severe polycythemia with hemoglobin at 22 g/dl and hematocrit at 74% which justified the use of a drain of 200 mm³. The patient has undergone an atrial switch according to the procedure of Senning–Mustard. Postoperative outcome was simple with a good operative result judged on clinical improvement and ultrasound control (Figs. 1 and 2).

Discussion

The transposition of the great arteries (TGA), with a prevalence of 0.213–0.442/1000 births, is the most frequently diagnosed CHD in the neonatal period [1].

It is also the noisiest CHD in this age and the most formidable. Since, it is the ultimate neonatal heart emergency. Indeed, the prognosis depends on early diagnosis and therapeutic management. Progress on antenatal screening for this type of heart disease has improved considerably their prognosis [2].

Another feature of TGA is its rapid evolution toward pulmonary hypertension and heart failure, making it spontaneously lethal. Indeed, over 50% of children with a simple TGA die during the first month of life. A small number survive to 6 months and about 90% of these patients die before 1 year. On average, the life expectancy of the unoperated simple TGA is 9 months [3,4].

Chantepie and al reported a preoperative mortality of 9.9% in neonates with TGA accessible to surgical correction, despite a correct management in specialized centers [5].

Our case report is unique in more ways than one.

First, it illustrates the exceptional survival of simple TGA. It has been diagnosed in our patient...
at the age of 11 years with polycythemia as the only complication.

Our review of the literature finds only two cases of simple TGA diagnosed in adulthood [6,7].

On the other hand, our patient’s pulmonary pressures remained low and the left ventricle (LV) preserved, which is unusual to our knowledge, seeing that the spontaneous evolution of transposition of the great vessels leads quickly to pulmonary hypertension and left ventricle dysfunction. That contra-indicates surgical correction in many cases or at least make it require prior left ventricle retraining.

Another peculiarity of our case is the primary success of surgical correction at this age, knowing that surgery in such pathology is mostly recommended in the neonatal period.

In the guidelines for the management of CHD in adulthood [8,9], no recommendation for TGA is reported. But the two cases of simple TGA in adulthood reported, have also undergone an atrial Switch using the technique of Senning–Mustard with a good surgical result [6,7].

The prognosis for our patient depends now on the risk of Mustard procedural complications, namely atrial arrhythmias, sinus failure and right ventricular dysfunction.

Conclusion

The transposition of the great arteries is an extreme neonatal emergency. Without an urgent treatment of this disease, early death is the rule. However, a prolonged natural survival seems miraculous but is possible, as evidenced by the observation we report.

References