

Ebstein' s anomaly: fetal presentation and neonatal outcome

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Objective: To identify predictors of poor outcome in neonates with Ebstein's anomaly (EB).

Background. EB is a rare congenital heart disease with variable neonatal presentation and outcome.

Patients and Methods. From January 2004 to January 2011, 16 newborns, 11 with a prenatal diagnosis, were referred to our centre.

Results: Among foetuses with EB 2 had a severe hydrops, 1 a tachyarrhythmia, 9 a cardio-thoracic ratio >0.6, 4 a reversed flow through the ductus arteriosus. At birth 14 patients had moderate to severe EB; 4 had congestive heart failure, 9 duct-dependent pulmonary circulation and needed PGE1 infusion, which restored an adequate pulmonary flow in 5 children.

3 patients with restrictive PFO and metabolic acidosis underwent an atrioseptostomy.

4 patients needed a BT shunt associated with plasty of the right atrium or closure of the tricuspid valve. There were 4 deaths, all occurred in infants with a prenatal diagnosis: 1 at birth, 2 postoperative and 1 sudden death 53 days after surgery. Patients with prenatal heart failure displayed a positive evolution under PGE1 and did not need any intervention.

At a median follow-up of 2 years, 2 patients had a Glenn palliation, the remaining 10 patients are symptoms free. The positive outcome was not related to the degree of tricuspid regurgitation, which showed the tendency to decrease with time, but with the area of functional right ventricle.

Conclusions: EB is a severe cardiac disease when it presents during the fetal or the neonatal period.

In this small series fetal hydrops was not related with perinatal exitus. Mortality was high and the outcome, not influenced by prenatal diagnosis, was related to the presence of functional right ventricle. All babies who survived after the neonatal period had a good outcome at medium term follow-up.