Prenatal diagnosis of isolated total anomalous pulmonary venous connection: a series of fourteen cases

Bonnet D. (1), Laux D. (1), Fermont L. (2), Le Bidois J. (1), Stirnemann J. (1), Bajolle F. (1), Boudjemline Y. (1), M3C-Necker, Université Paris Descartes, Sorbonne Paris Cité, Necker Enfants Malades, Paris, France (1); Institut de Puériculture de Paris, Paris, France (2)

Background: Prenatal diagnosis of total anomalous pulmonary venous connection (TAPVC) diagnosis is possible but remains a challenging issue in foetal echocardiography. The objective of this study was to determine accuracy and outcome of 14 foetuses diagnosed with isolated TAPVC.

Methods: We review our ten-year-experience of prenatal diagnosis of isolated TAPVC and describe the most relevant echocardiographic features. Medical records and echocardiographs of foetuses with a prenatal diagnosis of isolated TAPVC were retrospectively assessed. Special attention was paid to the foetal echocardiographic features motivating referral for expert echo scan.

Results: Expert foetal echocardiography identified 14 foetuses with isolated TAPVC. Prenatal diagnosis was made at a mean gestational age of 30 weeks (25 – 35 weeks). One woman was lost to follow-up before birth. There was one termination of pregnancy (TOP) for chromosomal abnormality. Two infants had a normal heart after delivery. Prenatal diagnosis of isolated TAPVC was confirmed in ten infants at postnatal assessment. All infants with confirmed TAPVC underwent corrective surgery. There was one in-hospital death after 1 ½ months due to respiratory failure. At the time of data collection, all patients were in good clinical condition without pulmonary hypertension or stenosis of pulmonary venous anastomosis after a mean follow-up of 30.5 months (1, 2-103, 8).

Conclusion: Foetal echocardiography can diagnose isolated TAPVC even in absence of complex congenital heart disease but remains challenging. The diagnosis has to be made by direct visualization of the pulmonary venous confluence behind the left atrium and absence of pulmonary veins connecting to the left atrium.