



SCIENTIFIC LETTERS

Right (atrial) appendage enlargement: Prenatal diagnosis and postnatal follow up[☆]



Dilatación de la orejuela derecha: diagnóstico prenatal y seguimiento postnatal

Dear Editor:

Right atrial appendage enlargement is an infrequent disease characterised by the enlargement of the appendage in the absence of any other cardiac lesions.¹ There are very few cases published in the literature, most of them in adults, and rarely diagnosed antenatally.² The age of onset ranges from the foetal period to adulthood. The clinical presentation in the paediatric population is variable, and most patients are asymptomatic and diagnosed based on incidental findings. It is also variable in affected fetuses, ranging from the absence of symptoms to foetal hydrops due to severe tricuspid regurgitation.³ It is essential that it be diagnosed due to its potential complications: hard-to-control tachyarrhythmias, thromboembolic events, dyspnoea, sudden death or heart palpitations. Its natural history is still not well understood, and this poses barriers to reaching a consensus on its management, so that treatment must be personalised based on symptoms and serial echocardiographic findings. Some authors propose watchful waiting in asymptomatic patients, while others recommend oral anticoagulant therapy or surgical resection depending on the disease course to prevent complications.²

We present two cases of right atrial appendage enlargement diagnosed prenatally, and their course to date.

Case 1

Patient aged 18 months in whom an aneurismal enlargement of the right atrium appendage was suspected prenatally, with echocardiography revealing an area of 4 cm² in the

four-chamber view at 31.6 weeks' gestational age, and no other family history of interest. The patient was born through elective caesarean delivery at 39 weeks' gestational age and admitted for monitoring. There were no complications during labour or the perinatal period. Treatment with acetylsalicylic acid (ASA) was initiated and is still ongoing due to persistent significant enlargement. The size of the atrium has decreased gradually, with an area of 8 cm² at 4 months of chronological age and 5 cm² in the last echocardiographic control at age 18 months, which revealed no intra-auricular thrombi and a normal right atrium morphology (Fig. 1). The serial echocardiograms have not detected arrhythmias or other complications, and the patient has remained asymptomatic and with normal findings in physical examination throughout the followup.

Case 2

Patient aged 15 months with right atrial appendage enlargement diagnosed prenatally at 20 weeks' gestational age, with an area of 1.6 cm² detected in the four-chamber view at 35 weeks' gestation. She was admitted to the hospital for twenty-four hours after an uncomplicated delivery at 40.1 weeks' gestation for observation and cardiac monitoring, and had no arrhythmia or other complications. The atrial appendage size has remained constant during the followup, with an area of 3.5 cm² observed in the four-chamber view. There has been no evidence of intra-auricular thrombi (Fig. 2). In this patient, given the absence of symptoms and complications, we adopted a watchful waiting approach and did not initiate treatment.

In the literature, the cases of idiopathic right atrial appendage enlargement that are diagnosed prenatally are exceptional. Since this is a rare disease that can present with significant complications, its early diagnosis allows an assessment of its course and informing the family more thoroughly.

The idiopathic enlargement of the right atrial appendage can be confused with other cardiopathies, such as Ebstein anomaly⁴ or, as occurred in our case during early prenatal tests, with a pericardial cyst, which was ruled out at a later time. The differential diagnosis with Ebstein anomaly is easy if there is evidence of normal insertion of the tricuspid valve.

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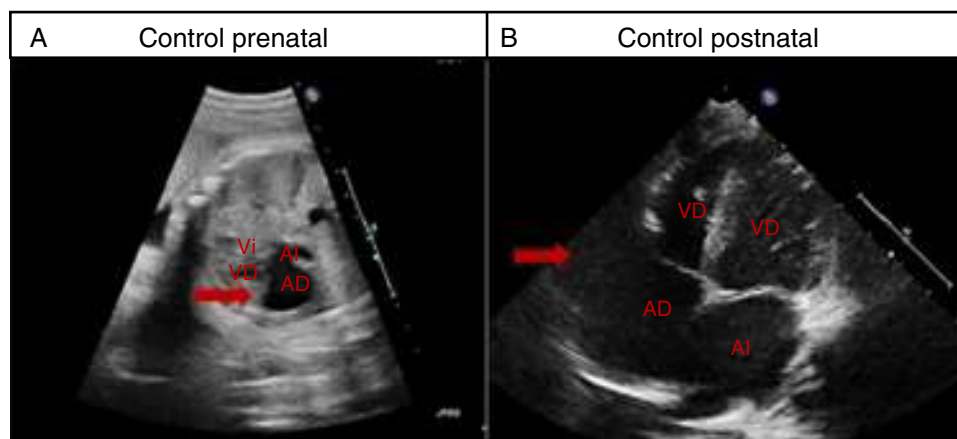


Figure 1 Case 1. Right atrial appendage enlargement. (A) Foetal ultrasound at 31 weeks' gestation, arrow points at right atrial enlargement. (B) Ultrasound: last postnatal followup at 18 months of age showing enlargement of right atrial appendage 5 cm² in diameter (arrow). AD, right atrium; AI, left atrium; VD, right ventricle; VI, left ventricle.

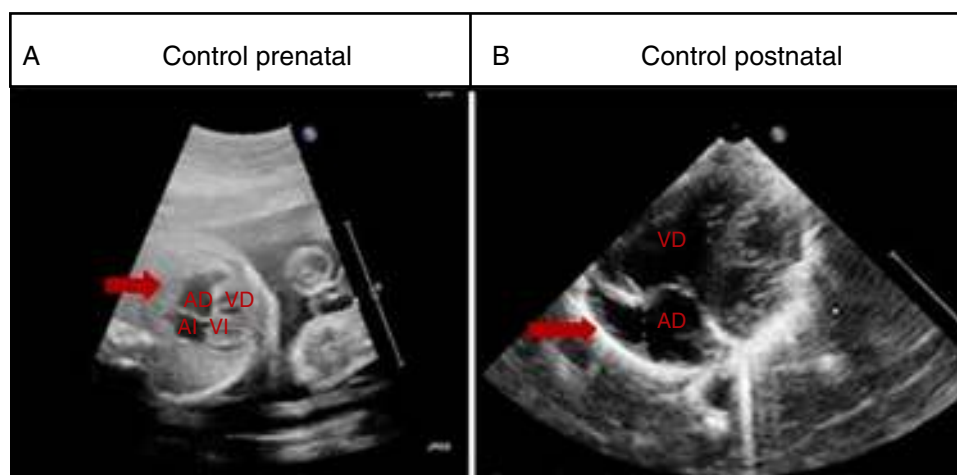


Figure 2 Case 2. Right atrial appendage enlargement. (A) Foetal ultrasound at 22 weeks' gestation, arrow points at right atrial enlargement. (B) Postnatal follow-up ultrasound at 1 year of life showing enlargement of the right atrial appendage 6 mm in diameter (arrow). AD, right atrium; AI, left atrium; VD, right ventricle; VI, left ventricle.

The postnatal diagnosis can be made by means of techniques such as transthoracic and transesophageal ultrasound, computed tomography and magnetic resonance imaging, although transthoracic echocardiography usually suffices for definitive diagnosis in the paediatric age group.⁵ Given the age of these patients and the optimal visualisation by means of transthoracic ultrasound to present, we have not needed other imaging tests that would require, at this time, the sedation or anaesthesia of the children, although they may be needed in the future.

Since the diagnosis was made prenatally in our patients and we performed serial echocardiograms, we could anticipate potential complications. We adopted a conservative approach at the outset due to the absence of symptoms, and in one case the extent of the enlargement eventually led to initiation of anticoagulant therapy, which is associated with few adverse effects. Depending on the course of the disease in each patient, and on a personalised basis, we will choose whether to maintain the conservative approach or use more invasive interventions, such as surgical excision.

The two cases presented here are exceptional because they were diagnosed before birth. This disease should be suspected when foetal echocardiography detects a right atrium-dependent dilation with no evidence of other possible causes. Its correct diagnosis makes it possible to provide appropriate follow-up care to prevent potential complications.

References

1. Rudski LG, Lai WW, Afilalo J, Hua L, Handschumacher MD, Chandrasekaran K, et al. Guidelines for the echocardiographic assessment of the right heart in adults: a report from the American Society of Echocardiography endorsed by the European Association of Echocardiography, a registered branch of the European Society of Cardiology, and the Canadian Society of Echocardiography. *J Am Soc Echocardiogr.* 2010;23: 685–713.
2. Hofmann SR, Heilmann A, Häusler HJ, Dähnert I, Kamin G, Lachmann R. Congenital idiopathic dilatation of the right

- atrium: antenatal appearance, postnatal management, long-term follow-up and possible pathomechanism. *Fetal Diagn Ther.* 2012;32:256–61.
3. Harder EE, Ohye RG, Knepp MD, Owens ST. Pediatric giant right atrial aneurysm: a case series and review of the literature. *Congenit Heart Dis.* 2014;9:E70–7.
 4. Patel V, Nanda NC, Rajdev S, Mehmood F, Velayudhan D, Vengala S, et al. Live/real time three-dimensional transthoracic echocardiographic assessment of Ebstein's anomaly. *Echocardiography.* 2005;22:847–54.
 5. Békássy SM, Kónya L. Idiopathic dilatation of the right atrium. A case report. *Scand J Thorac Cardiovasc Surg.* 1984;18:191–2.

Lidia Cardiel Valiente^{a,*}, Claudia Orden Rueda^b, Ariadna Ayerza Casas^a, Daniel Palanca Arias^a, Lorenzo Jiménez Montañés^a

^a *Unidad de Cardiología Pediátrica, Servicio de Pediatría, Hospital Universitario Miguel Servet, Zaragoza, Spain*

^b *Servicio de Pediatría, Hospital de Alcañiz, Alcañiz, Teruel, Spain*

*Corresponding author.

E-mail addresses: lidia_cardiel@hotmail.com, lidiacardielv@gmail.com (L. Cardiel Valiente).

Continuous flow peritoneal dialysis in a paediatric intensive care unit[☆]



Diálisis peritoneal de flujo continuo en una unidad de cuidados intensivos pediátricos

Dear Editor:

Acute kidney injury (AKI) is a severe complication in paediatric intensive care units (PICUs) whose incidence varies (2%–10%) depending on the population under study and the diagnostic criteria applied, and is found more frequently in neonates.¹ Different extracorporeal renal replacement therapies (RRTs) are available for its treatment, which are chosen based on the disease that led to AKI, the haemodynamic and respiratory status of the patient, and the resources available at the hospital.² Peritoneal dialysis (PD) is the first choice in patients with coagulation disorders or in whom venous access is difficult, situations that are common in paediatric patients. However, despite being an inexpensive option that does not require staff specifically trained in haemodialysis or much equipment, it is less effective than haemodialysis or other continuous RRTs in performing ultrafiltration (UF) and clearing solutes. In recent years, the technique of continuous flow peritoneal dialysis (CFPD) with a dual catheter system used in patients with chronic kidney injury in the 1960s and 70s has experienced a resurgence, with studies in the literature demonstrating higher clearances and UF compared to conventional PD, even in paediatric patients.^{3–6} Furthermore, the infusion of smaller volumes in the peritoneum results in improved haemodynamic and respiratory tolerability in critical patients.

The aim of this prospective, descriptive and observational study was to assess the feasibility, ultrafiltration efficacy and possible complications of dual-catheter CFPD in patients with AKI admitted to a PICU.

We included all patients admitted to a tertiary PICU between July 2013 and December 2014 that required PD for treatment of AKI after obtaining informed consent.

Description of the method

The procedure was carried out under mild sedoanalgesia. Using the Seldinger technique, two percutaneous Cook[®] PD catheters were inserted, of size 8.5Fr in infants and 11Fr in one school-aged child. One of the catheters was inserted approximately midway between the umbilicus and the left anterior superior iliac spine, and the other midway between the umbilicus and the right anterior superior iliac spine, positioning its tip inferiorly to the other catheter to facilitate intraperitoneal drainage. We used abdominal ultrasonography in some patients with small volumes of ascitic fluid to determine the best site for insertion. The first catheter was used to infuse the dialysis solution BicaVera[®] 1.5% glucose at a rate of 10 mL/kg/h with a continuous infusion pump, and the second catheter was connected to a urometer to measure the volume of dialysate drained via gravity on an hourly basis.

We collected and analysed data for the following clinical variables: age, sex, weight, underlying disease, Pediatric Risk of Mortality Score (PRISM), Kidney Disease Improving Global Outcomes Score (KDIGO), dialysate fill volume, glucose concentration of the dialysis solution, outflow volume, changes in fluid balance (FB) including diuresis and insensible losses, duration of treatment and associated complications.

We performed CFPD in five patients with clinical and laboratory signs of AKI in whom it was necessary to achieve a negative FB; four were in the post-operative period following surgery for congenital heart disease, and one had haemophagocytic syndrome secondary to infection by Epstein–Barr virus (EBV). Continuous flow peritoneal dialysis was chosen over other RRTs for the following reasons: its technical ease, as vascular cannulation is more difficult in young and unstable infants; its higher haemodynamic and respiratory tolerability, as lower fill volumes are required compared to intermittent dialysis techniques; and a lower risk of multiple transfusions and hypervolemia as well as of haemorrhage since heparin is not required, while it is in haemodialysis and other continuous RRTs.

The ages of the patients ranged from 21 days to 7 years. Four of the patients were male. We achieved a mean dialysis outflow rate of 1.0–7.1 mL/kg/h and a negative FB. We increased the dialysis inflow rate from 10 to 20 mL/kg/h in one patient, and subsequently observed a higher outflow rate (mean rate, 9.9 mL/kg/h). The criterion for discontinuation of CFPD was the improvement of

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