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ORIGINAL ARTICLE

Prenatal Diagnosis of Isolated Redundant Foramen Ovale: A Case Report

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Abstract Redundant foramen ovale (RFO) is defined as an abnormally redundant foramen ovale flap that extends at least halfway across the left atrium. The exact pathogenesis is unknown. Premature, isolated, in utero RFO is rare. The prevalence in general population is unknown as it may be easily ignored on routine fetal echocardiography. The reported frequency in fetuses referred for echocardiographic examination is 0.6–1.7%. It can cause right ventricular volume overload leading to fetal hydrops and subsequent cardiac failure. In such cases, prompt delivery depending on fetal gestational age may be instrumental for the survival of the neonate. 67% can develop cardiac arrhythmias which generally resolves at birth. The most common is premature atrial contractions though rarely, supraventricular tachycardia can also occur. RFO usually occurs in isolation but when associated with congenital heart disease, it carries a poor prognosis. Several series have reported redundant foramen ovale in echocardiographic and autopsy findings in children and adults but only few reports on the antenatal detection of isolated RFO has been described. We discuss two prenatal cases of isolated redundant foramen ovale diagnosed in the third trimester. They were followed up two weekly without development of any complications and delivered at term. After delivery, the infants had no history or symptoms of cardiac distress. Echocardiography showed a structurally normal heart. Hence, if there are no associated anomalies in a case of redundant foramen ovale, the prenatal management need not be altered.

Keywords Redundant foramen ovale · Fetal hydrops · Arrhythmia · Atrioventricular valve prolapse

Introduction

Formation of the inter-atrial septum in fetal life is complex. Early in development, a thin membranous septum primum appears in the fetal atrium separating the two atria. Later, the thicker septum secundum develops and grows along the side of septum primum. It has an oval opening in the middle third called the foramen ovale. This is an important communication allowing normal right-to-left atrial shunting during fetal life. It is guarded by the septum primum free flap which acts as a one-way valve with the flap bulging into the left atrium. Immediately after birth, the pulmonary venous return and the left atrial pressure start to rise and thereby, the foraminal flap of the septum primum adheres to the septum secundum and closes [1]. If the septum primum tissue grows extensively or its supporting tissue is scarce, the loose and wide septum primum flap (foraminal flap) becomes mobile [2]. If the enlarged flap extends as much as 50% into the left atrium in intrauterine life, this tissue is defined as redundant foramen ovale (atrial septal aneurysm, redundant septum primum flap, foramen ovale aneurysm) [3]. In the fetus, approximately 80% of the total volume of oxygenated blood that comes from the placenta passes through the umbilical vein and goes through the foramen ovale into the left atrium. Premature restriction of blood flow through the foramen ovale can cause right heart failure, resulting in fetal hydrops and eventually fetal death [1]. Isolated cases of redundant

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foramen ovale are often overlooked antenatally. In this article, we present two cases of isolated redundant foramen ovale that were followed up without development of complications.

Report of Cases

Two primigravidas were referred to us in their third trimester for a routine fetal ultrasound at 28 and 33 weeks respectively. Their first trimester screening and target ultrasound had no pathological findings.

Fetal sonography including M-mode, 2-dimensional, pulsed and color Doppler echocardiographic studies performed with GE Voluson E8, using transabdominal curvilinear transducers with frequency 2–5 MHz (4CA 2–5), and 1–5 MHz (C1-5-D), respectively (GE—General Electrics, Vienna, Austria). On evaluation of the four chamber view, the foramen ovale flap was stiff without the normal flapping motion. An abnormally redundant flap was seen which extended more than halfway across the left atrium (Fig. 1). On colour doppler, there was lateral displacement of flow around the foramen ovale flap from left atrium to left ventricle (Fig. 2). Also, a reversal of flow across the foramen ovale, i.e. left to right shunt was noted (Fig. 3). On M-mode, the flap excursion showed a uniphasic pattern instead of a biphasic one (Fig. 4). Pulmonary venous return and mitral orifice flow were not impaired. The intra-cardiac anatomy was otherwise normal without any signs of fetal hydrops or pleural or ascitic effusion. Fetal growth parameters were normal. The above ultrasonographic features were highly suggestive of the diagnosis of redundant foramen ovale.

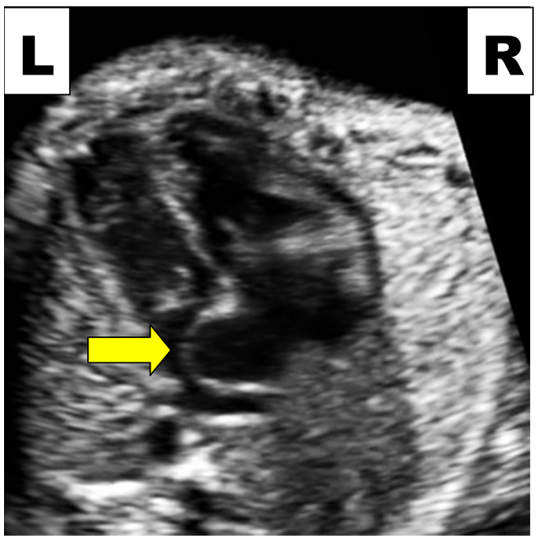

Fig. 1 Abnormally redundant foramen ovale flap extending more than halfway across the left atrium

After appropriate counseling, the cases were followed up after every two weeks to look for the development of complications. Finally, both patients delivered at term. Post-natally, both babies were evaluated and physical examination and echocardiography yielded no abnormal findings. They were discharged at days 3 and 6 of postnatal life. On follow-up 3 months later, the echocardiography was normal and babies are doing well.

Discussion

The foramen ovale is an opening in the septum secundum that is guarded by the septum primum flap. It is the most important inter-atrial communication in the fetal heart which provides right to left shunting of oxygenated blood across the atria and then into left ventricle supplying both cerebral and coronary vascular beds with adequately oxygenated blood [4]. Premature restriction of flow across the foramen ovale results in decreased blood flow into the left ventricle eventually leading to a reversal of flow across the foramen ovale. This leads to enlarged right heart that eventually causes fetal hydrops and intrauterine death depending on the time of onset and the severity of the redundancy. The incidence ranges from 0.2 to 1% depending on the study population [2]. Though the etiopathogenesis is unknown, it has been postulated that a true isolated primary RFO may be due to a localized developmental abnormality [5] or an in utero insult such as myocarditis [6].

Prenatal diagnosis is established from the four chamber view. On two-dimensional echocardiography, the normal foramen ovale in the fetus is seen as an echolucent area at the middle third of the interatrial septum. The overlying septum primum flap is freely mobile with unrestricted right to left flow. It normally bulges into the left atrium. The first prenatal diagnosis of foramen ovale obstruction was published 1982 by Hansmann and Redel [7].

ASA is a localized redundancy of the interatrial septum that bulges into the right and/or left atrium. It can be seen in up to 40% of fetuses referred for fetal echocardiography. There is a highly significant association between the presence of an RFO and an atrial arrhythmia in the fetus, most commonly premature atrial complexes (PAC) [8]. Papa et al. [9] reported ASA in 93 (7.6%) of 1223 fetuses, with 36% of them being associated with PACs. They concluded that the pressure exerted by the redundant septum primum on the atria had caused the ectopic rhythm. Post-natally, normalized pulmonary circulation causes the left atrial pressure to increase and thereby close of the foramen ovale flap, thus eliminating the cause for these PACs.

Cases of ASA have been reported with associated atrial septal defects, atrioventricular valve prolapse, or

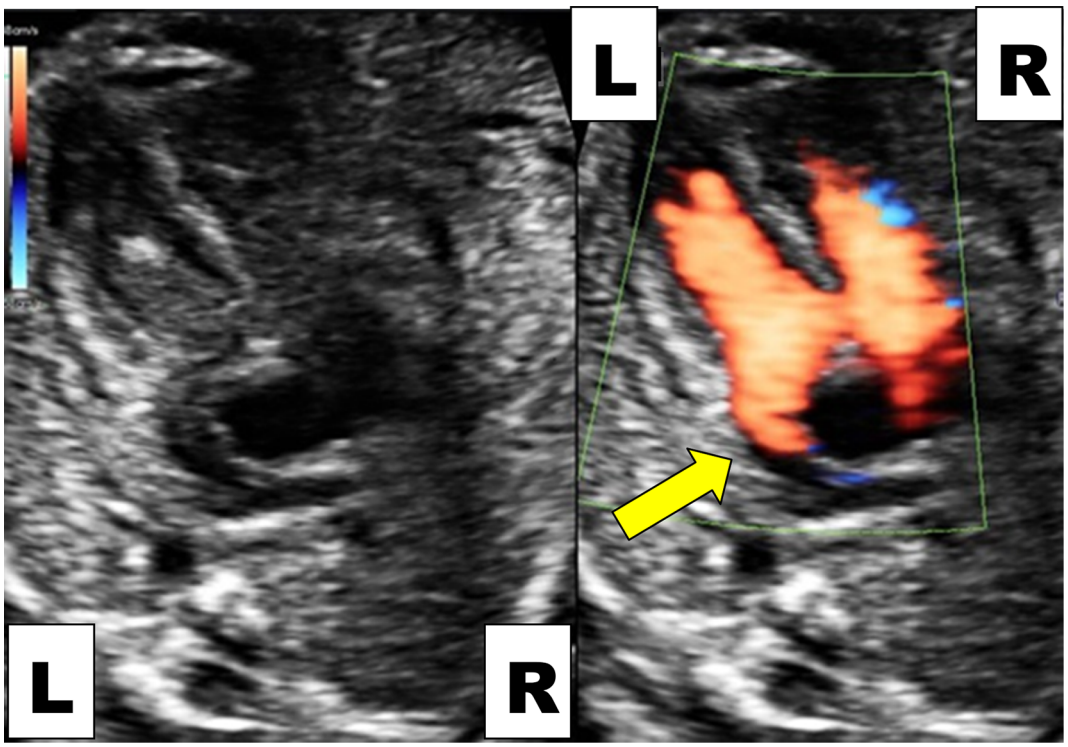

Fig. 2 Lateral displacement of flow around the foramen ovale flap from left atrium to left ventricle

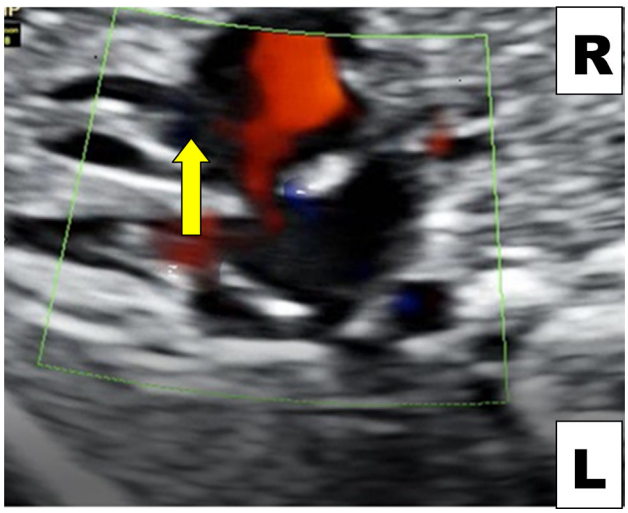

Fig. 3 Reversal of flow across the foramen ovale, from left to right

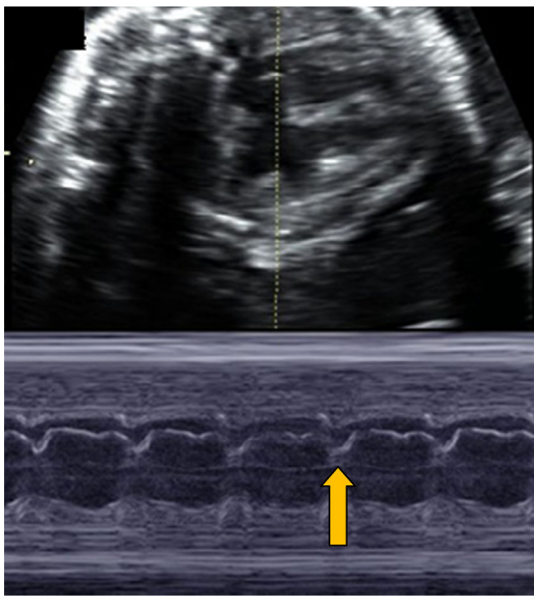

Fig. 4 Foramen ovale flap excursion showing uniphasic pattern

hypoplastic right heart variants [10]. In 2015, Heather et al. reported an unusual case of RFO in which the FO flap prolapsed into the mitral valve orifice. This resulted in a modifiable yet significant progressive cause of LV inflow obstruction, creating a potential for evolving hypoplastic left heart in later gestation. Due to the above concerns, the perinatal team electively delivered the fetus early at 34 weeks. Post-natally, the infant was clinically well without intervention, with rare premature atrial complexes noted on routine electrocardiogram [11].

Another concern is that of the development of fetal hydrops. When an isolated FO is the only underlying cause, complete reversal of the hydrops frequently ensues with timely delivery [6, 10].

Our two cases represent a situation in which FO obstruction manifested in later gestation. Progression to fetal hydrops and/or tricuspid regurgitation may have

occurred in the presence of earlier FO obstruction. A two weekly follow-up fetal echocardiography yielded no occurrence of complications and hence, both patients were delivered at term. Our cases were similar to that stated by Pineda et al. [12] and Dellinger [13], in that, despite the benign follow up, a close prenatal attention is needed for development of complications.

Conclusion

The foramen ovale flap should be evaluated more critically in the four chamber view so that all the cases of isolated redundant foramen ovale are picked up. The unexpected presence of such a finding should warrant further search for additional cardiac and extracardiac abnormalities, which may alter the prognosis. In isolated cases, a monthly follow up is advised to look for development of arrhythmias or fetal hydrops. A closer surveillance is advocated in the advent of developing signs of obstruction. Arrhythmias are self resolving in these cases. Prompt delivery is a life-saving measure for fetal hydrops with adequate gestational age and lung maturity.

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