

# Left atrial isomerism associated with aneurysmal enlargement of right atrial appendage: A case report with literature review

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## Abstract

We present a prenatally diagnosed case of heterotaxy syndrome (HS) in which left atrial isomerism (LAI) was associated with an aneurysmal enlargement of the right atrial appendage (RAA). Although LAI is usually associated with complex cardiac and extracardiac anomalies, the association of LAI and right atrial appendage aneurysm (RAAA) is exceptional. Congenital RAAA itself is an idiopathic, very rare cardiac anomaly characterized by the enlargement of the appendage in the absence of any other cardiac or extra-cardiac defect. The prognosis of the heterotaxy is poor with associated major cardiac malformations and even cases with minor cardiac anomalies are at risk postnatally for complications like biliary atresia, intestinal rotational abnormalities, and immune disorders. In this case, the prenatal diagnosis of the isomerism was mainly based on the abnormalities of caval veins. Although no typical complex cardiac anomaly was present, the HS was associated with biliary atresia, polysplenia, and malrotation of the gut. Associated RAAA further imposed an additional risk of complications such as tachyarrhythmias, thromboembolic events, and aneurysmal rupture.

**Key words:** Aneurysm; atrial appendage; heterotaxy; isomerism; polysplenia

## Introduction

HS also is known as situs ambiguous is a rare malformation syndrome, defined as an abnormal arrangement of viscera across the left/right axis primarily induced by disorders of laterality determination during early embryonic development.<sup>[1,2]</sup> HS differs from situs solitus and inversus and, in general, is classified into right atrial and left atrial isomerism.<sup>[3]</sup> Classical prenatal finding in both varieties is

viscerocardiac heterotaxy, complex cardiac malformations, and anomalies of the caval veins.<sup>[4]</sup> In most cases, fetal HS coexists with a varied spectrum of cardiac and extracardiac anomalies.<sup>[5]</sup> However, the association of the complex syndrome with RAAA is atypical and notable. The RAAA is a very rare anomaly, seldom detected, and reported prenatally and postnatally.<sup>[6]</sup>

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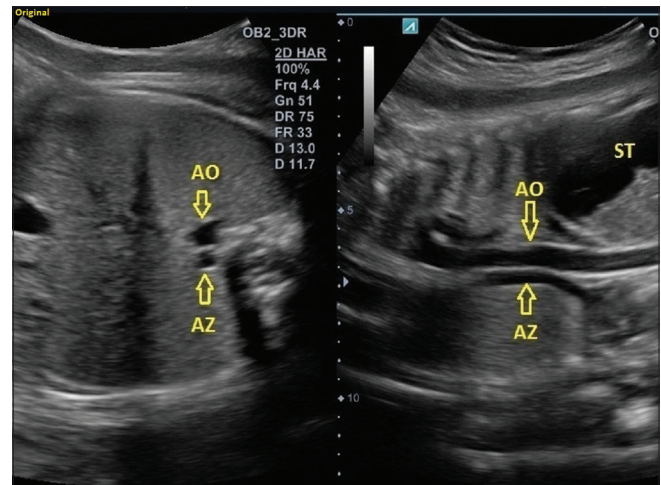
## Case Report

A routine fetal sonographic scan of a 29-year-old second gravida showed a singleton fetus of 27 weeks gestation. Fetal abdomen scans revealed dilated azygous vein lying posterior to the abdominal aorta [Figure 1]. In a bicaval view and three-vessel view, the azygous vein was visualized draining into the superior vena cava (SVC) [Figure 2]. The inferior vena cava (IVC) was interrupted. The hepatic veins were directly opening in the right atrium [Figure 3]. The inferior part of the IVC was malpositioned and was located on the left side of the spine, anterior to the descending aorta [Figure 4]. The portal sinus was not visualized. The four-chamber view revealed enlarged right atrium, aneurysmal dilation of the RAA, a dilated coronary sinus, azygous vein placed posterior to the descending aorta [Figure 5], and a small interventricular septal defect. The three-vessel view showed pulmonary trunk bifurcation, persistent left superior vena cava (PLSVC), a dilated SVC, and azygous vein [Figure 5]. The kinetic RAAA showed the inconsistent configuration and was lying anterior and right of the right atrium, best visualized in a bicaval view [Figure 6]. The aneurysm further got enlarged with advancing gestation [Figure 7]. Fetal abdomen scans at 32 weeks revealed a midline liver [Figure 8]. The gallbladder was not visualized. Multiple echo-poor circumscribed areas, along the greater curvature of the left-sided stomach, arouse suspicion of multiple splenules [Figure 8]. Serial scans disclosed intrauterine growth restriction. However, no complications related to the heterotaxy such as arrhythmia or hydrops developed till 37 weeks of gravidity. Based on sonographic findings, a diagnosis of LAI associated with RAAA was made and the possibility of coexisting extracardiac anomalies like polysplenia, gut malrotation, biliary atresia was explained to the couple. The family denied for a fetal MRI.

Cesarean section was done at 38 weeks of gestation. Postnatal investigations confirmed all major prenatal ultrasound findings including suspected biliary atresia and polysplenia, and additionally, gut malrotation was also diagnosed.

## Discussion

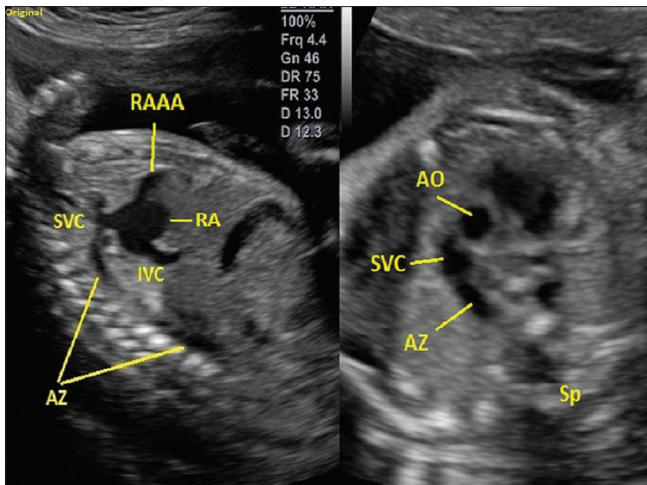
Left atrial isomerism and right atrial isomerism are conditions in which structures are bilateral within the thoracoabdominal cavity with morphologic left and morphologic right characteristics, respectively.<sup>[7]</sup> Typical arrangement of the organs associated with classic LAI is bilateral hyparterial bronchial branching pattern, bilateral morphological left atrial appendages, and polysplenia.<sup>[1,4]</sup> Main characteristic features of RAI are bilateral eparterial bronchial branching pattern, bilateral morphological right atrial appendages, and asplenia.<sup>[1,4]</sup> However, an exception to these general hallmarks is not infrequent as the features of classic isomerism were found breached in >20% of cases.<sup>[8]</sup>



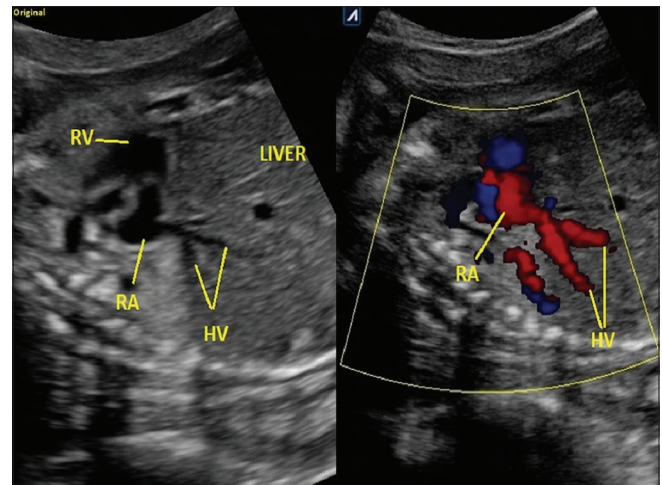
**Figure 1:** Double vessel sign: transverse and sagittal scans of the fetal abdomen at 27 week of gestation. The dilated azygous vein (AZ) is located side-by-side and slightly posterior to the descending aorta (AO) along the spine. ST: Stomach bubble

Since HS presents with considerable combinations of organ abnormalities and arrangement, prenatal sonographic diagnosis of the condition based on cardiac and extracardiac lesions is not possible. Furthermore, many anomalies such as biliary atresia, malrotation of gut, the bilobed or trilobed lung cannot be detected precisely by antenatal sonography. Determination of the morphology of the atrial appendages and, hence, defining atrial situs during fetal echocardiography<sup>[9]</sup> and cross-sectional imaging would provide the most accurate diagnosis of HS. The typical signs in cases of classic left isomerism include the bilateral sickle-shaped atrial appendages and the left configured atrial wall.<sup>[9,10]</sup> The classic right isomerism is characterized by bilateral pyramidal appendages and the right configured atrial wall.<sup>[9,10]</sup> Although the prenatal diagnosis of heterotaxy based upon the configuration of the atrial appendages has been reported,<sup>[11]</sup> it is difficult to assess these appendages because of their small size and inconsistent configuration.<sup>[9,10]</sup> Moreover, the atrial appendage morphology is not always indicative of bronchopulmonary or abdominal situs. Discordance between atrial appendage arrangement, splenic status, and bronchopulmonary branching was identified in a fair number of patients of heterotaxy postnatally.<sup>[8]</sup> Therefore, *in vivo*, atrial situs determination is mainly based on the general pattern of the associated cardiovascular and noncardiovascular findings than on the actual evaluation of the atrial morphology.<sup>[8]</sup> Interrupted IVC with azygous continuation into the SVC is a specific prenatal sonographic marker of LAI and was seen in 89.2%<sup>[5]</sup> to 100%<sup>[10]</sup> cases, while the juxtaposition of the aorta and the IVC is the main diagnostic feature of RAI in utero and was present in 81.5% fetuses.<sup>[4]</sup> Fetal heart block detected in the first trimester is also indicative of LAI and was observed in 59.3% of cases.<sup>[4,5]</sup> Discontinuity of the IVC is due to the absence of hepatic part of the vessel.<sup>[4,12]</sup> Systemic venous flow beyond the suprarenal part of the caval vein

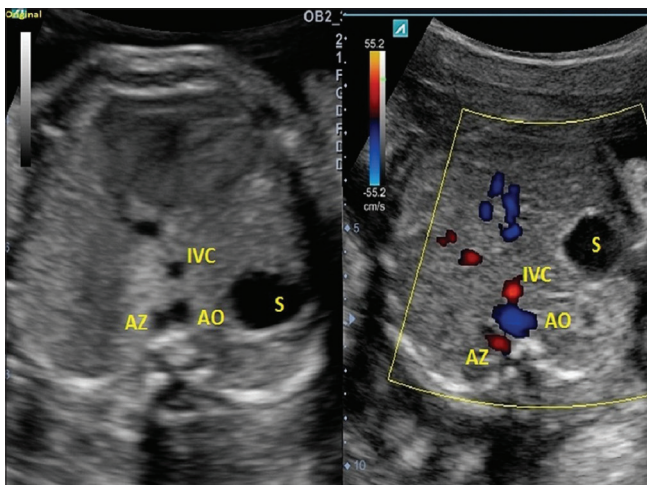




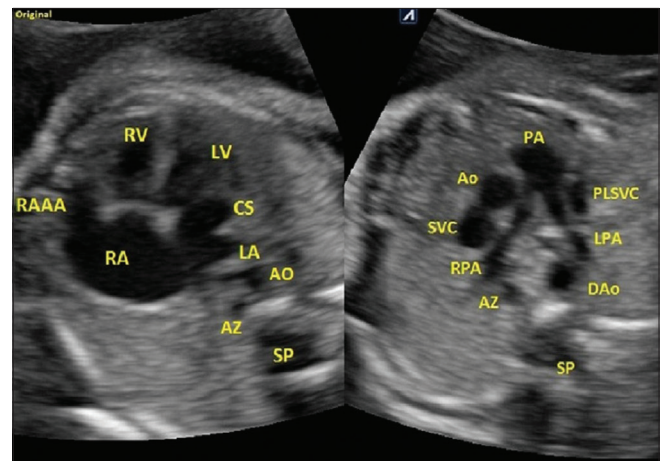
**Figure 2:** Bicaval and three-vessel view: dilated azygos vein (AZ) is draining into the SVC. Note the suprahepatic portion of the interrupted IVC and right atrial appendage aneurysm (RAAA), depicting exceptional association of heterotaxy and the aneurysm



**Figure 3:** Grey scale and color Doppler scans: The hepatic veins (HV) are draining directly into the right atrium (RA) in the absence of hepatic part of the IVC. RV: Right ventricle



**Figure 4:** Grey scale and color Doppler scans of fetal abdomen showing double vessel sign and abnormal position of the inferior part of the IVC. Portal sinus is not visualized. AZ: Azygos. AO: Descending aorta, S: Stomach bubble

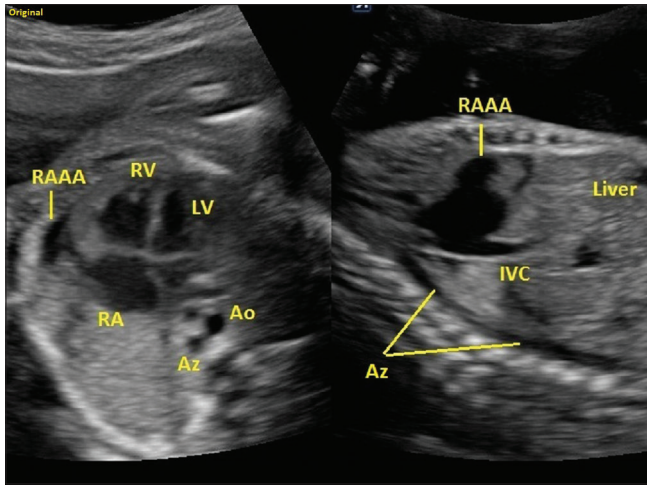


**Figure 5:** Four-chamber view (left panel) showing right atrial appendage aneurysm (RAAA) and dilated coronary sinus (CS). Three-vessel view (right panel) demonstrating persistent left superior vena cava (PLSVC) and dilated right SVC. RV: Right ventricle, LV: Left ventricle, RA: Right atrium, PA: Pulmonary trunk with right and left branches (RPA, LPA). AZ: Azygos vein. Ao and DAo: Ascending and descending aorta

is drained by dilated azygos or hemiazygos vein, which empty either into the right-sided [Figure 2] or an accessory left-sided SVC.<sup>[13]</sup> Suprahepatic part of the IVC opens at the bottom of the right atrium via a separate opening [Figure 2] and its inferior part remain malpositioned to the left of the spine [Figure 4]. Interruption of the IVC with azygos continuation can be detected prenatally as a “double vessel” sign<sup>[14]</sup> [Figure 1]. The sign demonstrates the azygos vein posterior to the descending aorta with the flow of blood in the opposite direction [Figure 4]. Diagnosis of caval vein abnormalities is of utmost importance as the grave extracardiac abnormalities, related to the heterotaxy, may be present even in the presence of a structurally normal heart.

PLSVC is seen in 4.4% of cases of heterotaxy.<sup>[2]</sup> In the vast majority, it is accompanied by a normal right-sided SVC,

termed SVC duplication.<sup>[2]</sup> In the three-vessel view, the PLSVC presents as a supernumerary vessel to the left of the pulmonary trunk [Figure 5]. Its drainage is variable and can be to the right atrium via the oblique vein of Marshall, the coronary sinus or the left atrium.<sup>[2]</sup> When it drains into the left atrium, a bubble-like coronary sinus is seen lying at the inferior-posterior aspect of the morphologic left atrium<sup>[9]</sup> [Figure 5]. The discontinuity and malposition of the IVC, as well as the duplication of the SVC, are based on disorders of the left-right determination of the paired primitive veins in early embryonic development.<sup>[2]</sup> The caval anomalies are frequently associated with portal vein abnormalities. The vein can be absent, atretic, hypoplastic, or malpositioned.<sup>[8]</sup> The malpositioned preduodenal portal vein is frequently associated with polysplenia syndrome

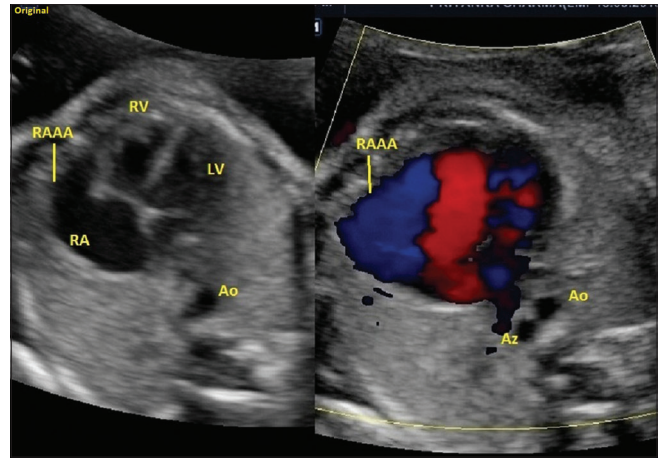


**Figure 6:** Four-chamber view (left panel) and bicaval view (right panel) showing right atrial appendage aneurysm (RAAA) with double vessel sign and interrupted IVC depicting coexistence of heterotaxy and RAAA. Az: Azygous vein, Ao: Descending aorta, IVC: Suprahepatic part of the inferior vena cava

and midline liver.<sup>[15]</sup> The anomaly is surgically important because it may produce pressure symptoms on the duodenum and bile duct.<sup>[16]</sup> The preduodenal ascension of the portal vein is an important radiological sign to diagnose polysplenia syndrome postnatally.<sup>[12]</sup>

Fetuses with typical LAI are inherently at risk for rhythm disturbances such as sinus bradycardia or complete atrioventricular (AV) block. Sinus bradycardia occurs due to the hypoplasia or absence of the sinus node,<sup>[17,18]</sup> whereas a complete AV block is a result of the discontinuity between the AV node and His-Purkinje system.<sup>[17,18]</sup> The sinus node is expected to be absent bilaterally in left isomerism. Dual sinus nodes and dual AV nodes in right isomerism may cause supraventricular tachycardia.<sup>[18,19]</sup> However, bilateral sinus nodes were observed in only 54% of cases with right isomerism, and bilateral absence of sinus nodes was documented in only 25% of cases with left isomerism.<sup>[19]</sup> The findings suggest that the sinus node is not a morphologically right-sided structure, and its presence, therefore, is not consistently related to the sidedness of the atria.<sup>[19]</sup> Thus, even in classical cases of left isomerism, heart block is not observed in all cases. Nevertheless, fetal heart block detected in the first trimester strongly suggests the presence of LAI, as immune-mediated heartblock does not develop before the antibodies start to cross the placenta in the early second trimester.<sup>[20]</sup> Fetuses with heterotaxy syndrome and bradycardia, either sinus or due to AV block should be closely followed-up during pregnancy to monitor the development of cardiac dysfunction and/or hydrops. Fetuses with normal cardiac function and without hydrops can be delivered near term irrespective of the low ventricular rate.<sup>[21]</sup>

Fetal HS coexists with a wide variety of complex cardiac anomalies, with abnormalities of the AV valves being



**Figure 7:** Grey scale and color Doppler axial four-chamber views at 32 weeks of gravidity: showing right atrial appendage aneurysm (RAAA). Color Doppler showed kinetic nature of the aneurysm. Az: Azygous, Ao: Descending aorta, RV: Right ventricle, LV: Left ventricle

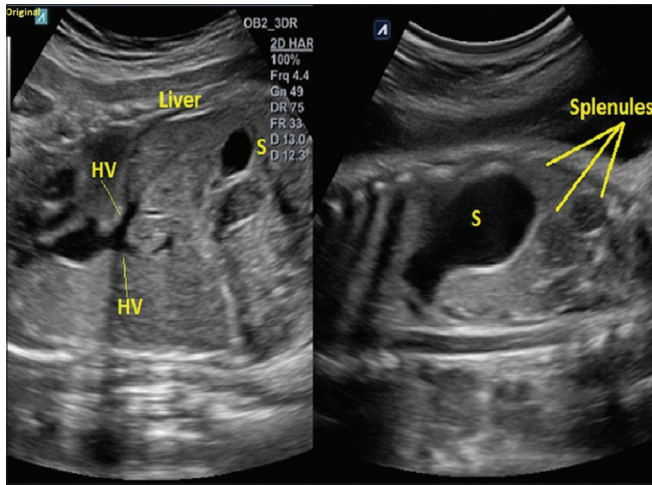
among the most common.<sup>[5]</sup> Other major cardiac defects associated with LAI are obstructive lesions of the right outflow tract, transposition or malposition of the great arteries, and double outlet right ventricle.<sup>[5]</sup> LAI, in general, have less severe cardiac malformations than those with RAI and may even present with the only interruption of the IVC. However, interrupted IVC may be seen in fetuses not affected by isomerism.<sup>[22]</sup>

In this case, LAI was associated with PLSVC, dilated left atrial coronary sinus, and small atrioventricular septal defect but no major typical cardiac anomaly was identified. However, the heterotaxy was associated with an exceptionally rare anomaly where there was aneurysmal dilation of RAA [Figures 6 and 7]. Usually, the atrial appendages are not visualized in fetal echocardiography due to their small size, uncertain configuration, and location.<sup>[9]</sup> The dilated RAA showed inconsistent configuration, lying anterior and right of the right atrium, with its apex pointing anteriorly [Figure 6]. Color Doppler manifested regurgitation of the blood from the aneurysm into the right atrium during atrial systole, mimicking tricuspid regurgitation. RAA is the most infrequent location for cardiac aneurysmal dilation.<sup>[23]</sup> The entity is an idiopathic, isolated, very rare cardiac anomaly with only a few reported cases in the literature.<sup>[6]</sup> Differential diagnoses of the RAAA include Ebstein's anomaly,<sup>[24]</sup> pericardial cyst,<sup>[25]</sup> and pleural effusion.

Associated major cardiac anomalies are the main determinant in anticipating the outcome for children affected by heterotaxy. However, even those presenting with isolated defects are at risk of intestinal malrotation, biliary atresia, and immune disorders due to polysplenia/asplenia.

Biliary atresia was diagnosed postnatally in 10.4%<sup>[26]</sup> of neonates with LAI. Prenatal nonvisualization of the fetal





**Figure 8:** Coronal scans of fetal abdomen: at 32 weeks gestation (left panel) showing mid-line liver, hepatic veins (HV) are seen draining into the right atrium. At 37 weeks (right panel): Multiple echo-poor widely spaced splenules suggesting polysplenia. S-stomach

gallbladder or a small gallbladder may be associated with biliary atresia.<sup>[27,28]</sup> However, the diagnosis of the disorder is not easy because of the highly variable gallbladder morphology. Estimation of digestive enzyme gamma-glutamyl transpeptidase in amniotic fluid<sup>[29]</sup> or fetal blood<sup>[30]</sup> was reported as useful. MRI can improve the prenatal detection and can differentiate between the atresia and choledochal cyst.<sup>[31]</sup> Biliary atresia is a progressive disease with jaundice and is the most common indication for Kasai's portoenterostomy or liver transplantation.<sup>[32]</sup>

Although polysplenia is the most common finding in left isomerism [Figure 8], asplenia or a normal right-sided or left-sided spleen may also be present.<sup>[33]</sup> Since the presence of one or multiple spleens does not adequately reflect splenic function, antibiotic prophylaxis and adequate immunization are required to prevent sepsis in a patient suffering from the heterotaxy.<sup>[34,35]</sup>

Anomalies of midgut derivatives include nonrotation, incomplete rotation, and the rare reversed complete or incomplete rotation.<sup>[10]</sup> The malrotation and malfixation of the bowel are associated with the portal vein and the superior mesenteric vessels anomalies. MRI is useful for the prenatal diagnosis of bowel malposition.<sup>[35]</sup> Diagnosis of the condition is important as the abnormality may lead to midgut volvulus.

The fetuses suspected of being affected by heterotaxy should be referred for a fetal or neonatal MRI to exclude associated extra-cardiac defects which may strongly affect the long-term outcome.<sup>[36,37]</sup> Similarly, a prenatally diagnosed case of RAAA needs serial echocardiograms and postnatal follow-up because of the anticipated potential complications such as arrhythmia, thrombosis, pulmonary embolism, and rupture of the aneurysm.

Postnatal oral anticoagulant therapy or surgical resection of the aneurysm may be required to prevent these complications.<sup>[38]</sup>

Precise prenatal diagnosis of HS and associated anomalies not only helps in proper parental counseling, but it is crucial for prenatal and postnatal management with an interdisciplinary approach.

#### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

#### Conflicts of interest

There are no conflicts of interest.

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