

CONGENITAL MINI-FOCUS ISSUE

ADVANCED

CASE REPORT: CLINICAL CASE

Senning Procedure for Physiological Atrial Inversion With Left Atrial Isomerism



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ABSTRACT

We present the case of an infant with left atrial isomerism with complex pulmonary and systemic venous connections that resulted in physiological parallel circulation in the setting of ventriculoarterial concordance who was surgically treated using the Senning procedure. This case highlights a rare cause for cyanosis due to poor mixing from a parallel circulation and an issue with nomenclature. (**Level of Difficulty: Advanced.**)

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HISTORY OF PRESENTATION

A baby boy was emergently delivered by cesarean section at 35 weeks of gestation secondary to persistent fetal bradycardia to a 29-year-old, gravida 2 para 2 mother. Birth weight was 2.1 kg, and Apgar scores were 4, 6, and 8 at 1, 5, and 10 min, respectively. Pregnancy was complicated by gestational diabetes and urinary tract infection, but there was no family history or a prior fetal diagnosis of congenital heart disease. The infant was intubated immediately after

birth for cyanosis and respiratory distress. His physical examination at birth was notable for severe bradycardia (heart rate 50 to 60 beats/min), cyanosis (oxygen saturation 70%), and no appreciable murmurs. Breath sounds were diminished in both lung fields. Examination of other systems were reportedly unremarkable.

DIFFERENTIAL DIAGNOSIS

The complex anatomy of the present case is shown in **Figure 1**. The initial differential diagnosis included persistent pulmonary hypertension of the newborn, respiratory distress syndrome secondary to prematurity, congenital diaphragmatic hernia (CDH), and cyanotic congenital heart disease such as complete transposition of the great arteries (TGA) and obstructed total anomalous pulmonary venous return.

INVESTIGATIONS AND MANAGEMENT

Chest radiography was diagnostic for a right CDH with the intestinal contents in the right side of the chest, with complete opacification of the left lung (**Figure 2**). Electrocardiography demonstrated

LEARNING OBJECTIVES

- Multimodality imaging, including chest radiography, echocardiography, angiography, and computed tomographic angiography, is critical to define the segmental anatomy precisely in heterotaxy syndrome.
- Venoatrial connection abnormalities can have a profound impact on clinical presentation and management.
- The Senning procedure can be successfully performed in isolated physiological atrial inversion with LA isomerism.

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Informed consent was obtained for this case.

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junctional bradycardia and a markedly prolonged corrected QT interval of 600 ms (Figure 3). Echocardiography showed a midline liver, levocardia, possible bilateral left atrial (LA) appendages, unobstructed total anomalous pulmonary venous drainage to the right-sided morphological left atrium, the entire systemic venous drainage through a left-sided superior vena cava (SVC) and an uninterrupted inferior vena cava, both of which coursed to the left of the spine (Figure 4) before draining directly into the left-sided morphological left atrium. The left-sided systemic venous atrium connected to a left-sided morphological left ventricle (d-loop ventricular mass), and the right-sided pulmonary venous atrium connected to a right-sided morphological right ventricle, resulting in parallel circulation. The ventriculoarterial connections were concordant, and the great arteries were normally related. In addition, there was a small patent foramen ovale with bidirectional shunting, an intact ventricular septum, and a large ductus arteriosus with predominantly left-to-right shunting. Isomerism of the LA appendages was suspected with complex segmental anatomy. The sequential segmental anatomy would be LA isomerism, ambiguous biventricular atrioventricular connection with d-looping, and ventriculoarterial concordance and would seem most consistent with {A, D, S}. Computed tomographic angiography confirmed the anatomy, including the venous connections to the atria, morphologically bilateral LA appendages (Figure 5), and bilateral morphological left bronchi (Figure 6). Additional findings included left ventricular noncompaction (Figure 7), but biventricular systolic function was normal.

MEDICAL HISTORY AND MANAGEMENT BEFORE SENNING

The infant had severe persistent cyanosis after birth that was attributed to CDH, associated pulmonary hypertension of the newborn, and parallel circulation because of the abnormally discordant systemic and pulmonary venous drainage. An urgent balloon atrial septostomy was performed, and he underwent successful surgical repair of the CDH on day 2 of life. While recuperating from CDH surgery, cyanosis persisted, and a 3.5-mm central aortopulmonary shunt was placed and the ductus arteriosus was ligated at 1 week of age. Following surgical shunt placement, the infant initially had persistent cyanosis, but chest radiography showed prominent pulmonary vascular markings. Cardiac catheterization was performed that affirmed the previously described anatomy, including especially the systemic venous drainage anatomy

with a left-sided SVC and the hepatic veins that joined the uninterrupted inferior vena cava before draining directly into the left-sided atrium (Figure 8). Hemodynamic status at cardiac catheterization was as follows: mean right atrial pressure 11 mm Hg, pulmonary artery pressure 43/26 mm Hg (mean 25 mm Hg), pulmonary blood flow 7.35 l/min/m², systemic blood flow 3.13 l/min/m², and pulmonary blood flow/systemic blood flow ratio 2.35:1, and yet his systemic saturation was only 76%, consistent with the predicted parallel circulatory physiology.

SURGICAL PROCEDURE (SENNING)

At 3.5 months of age, the infant underwent a modified Senning procedure, take-down of the central shunt, and placement of an epicardial dual-chamber pacemaker. In this case, the systemic and pulmonary venous return was “transposed,” resulting in “physiological atrial inversion” and creating a circulatory pattern resembling classical d-TGA. The rationale for the Senning procedure was to create a physiologically normal series circulation by redirecting the systemic and pulmonary venous

ABBREVIATIONS AND ACRONYMS

- CDH = congenital diaphragmatic hernia
- SVC = superior vena cava
- LA = left atrial
- TGA = transposition of the great arteries

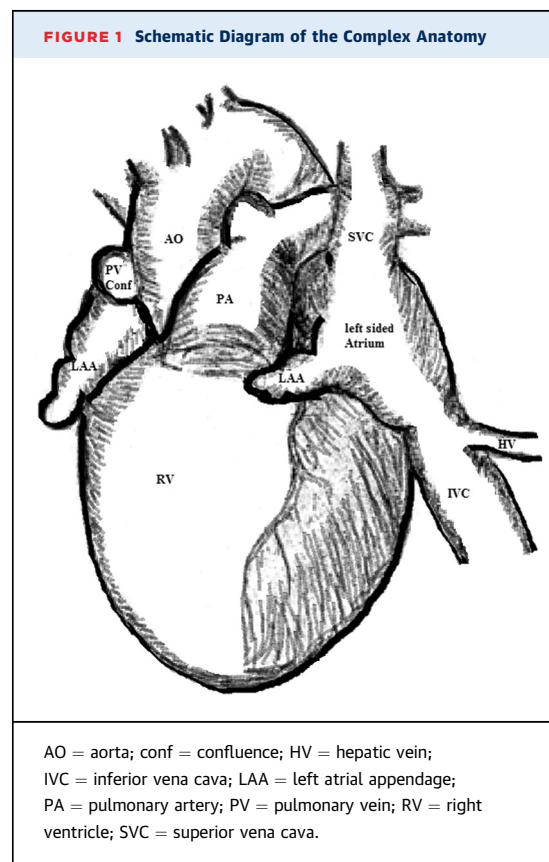
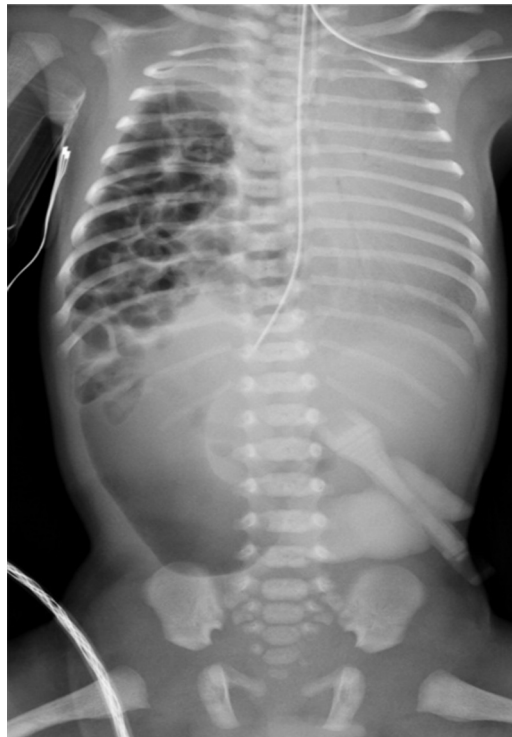


FIGURE 2 Chest Radiograph at Birth Showing Right-Sided Congenital Diaphragmatic Hernia



return to the appropriate ventricles using intra-atrial baffles, and the left ventricle remained as a systemic ventricle (1).

EXTRACARDIAC FINDINGS

The infant's chromosomal microarray was negative for long-QT syndrome but was positive for a mutation (c.663T>A, p.Tyr221) in the *DSC2* gene, which has been associated with arrhythmogenic right ventricular cardiomyopathy but not reported with LA isomerism. The infant also had the following extracardiac anomalies: intestinal malrotation, bilateral inguinal hernias, a dual collecting system and right-sided hydronephrosis, nephrolithiasis, and multisutural craniosynostosis. He underwent multiple noncardiac procedures, including a tracheostomy for ventilator dependency, Ladd's procedure, herniorrhaphy, Nissen fundoplication, gastrostomy tube placement, and open cranial vault remodeling surgery for craniosynostosis. He was discharged from the hospital at 4.5 months of age.

FOLLOW-UP

The infant's tracheostomy was successfully decannulated at 18 months of age. Despite his complex

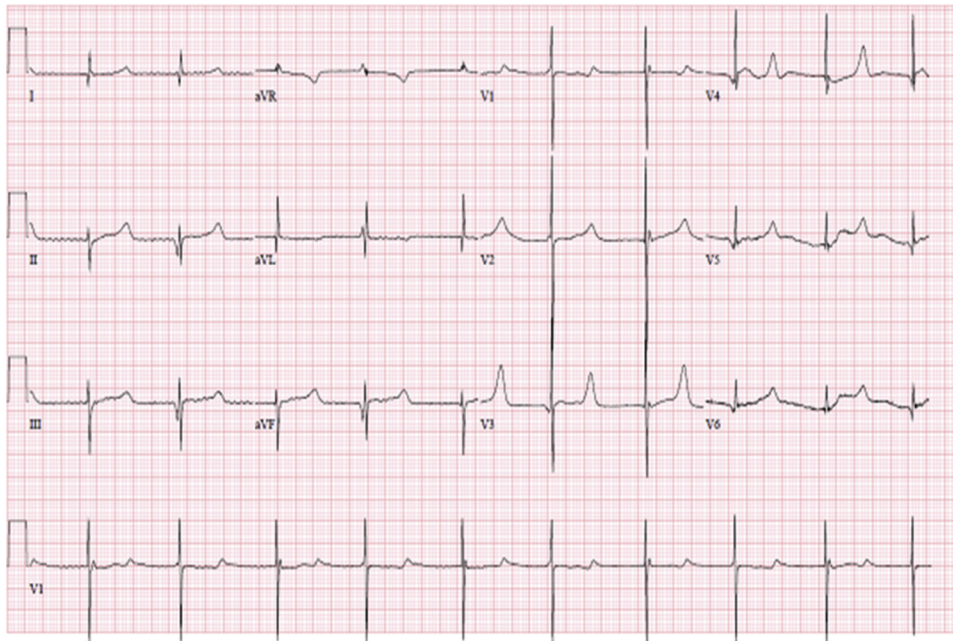
multisystem issues, he has been clinically doing well and thriving. Echocardiography at 32 months of age showed no obstruction of the intra-atrial systemic or pulmonary venous baffles (Figure 9), normal biventricular systolic function, and minimal atrioventricular regurgitation, without evidence of pulmonary hypertension.

DISCUSSION

Heterotaxy syndrome is defined as an abnormality of laterality in which the thoracoabdominal organs demonstrate abnormal arrangement across the left-right axis of the body (2). The morphology of the atria and in particular the atrial appendages often allow determination of "sidedness." This can be determined by echocardiography, but sometimes other imaging modalities such as angiography and computed tomographic angiography are essential for diagnosis, as they also allow clearer delineation of the arrangement of thoracoabdominal organs. LA isomerism or bilateral left-sidedness refers to duplication of the left-sided structures with features such as bilateral LA appendages, persistent left SVC, pulmonary veins connecting bilaterally to the atria (60%), interruption of the inferior vena cava, and additional complex intracardiac defects (3). Right atrial isomerism or bilateral right-sidedness can be associated with bilateral SVC, TGA, pulmonary artery stenosis or atresia, and an atrioventricular septal defect. Evaluation of the heart in patients with heterotaxy syndrome requires a complete description of both atrial appendages and the venoatrial, atrioventricular, and ventriculoarterial connections of the cardiac segments.

In our patient, physiologically discordant atrioventricular connections due to pulmonary venous drainage were to the right-sided morphological left atrium and systemic venous drainage to the left-sided morphological left atrium, but concordant ventriculoarterial connections constituted a circulation that resembled classical d-TGA (Figure 1). This case demonstrates the complexity of ascribing a nomenclature label to a rare complex abnormal connection, be it descriptive or numeric. The old adage of describing the morphological structures as it exists holds true. In this case, it predicts the true hemodynamic physiology of a parallel circulation without the usual ventriculoarterial discordance. According to morphological description, this patient had transposed systemic and pulmonary veins, resulting in a parallel circulation akin to d-TGA. Its morphological name is as yet undescribed, and perhaps it can be best described as physiological venoatrial discordance in

FIGURE 3 Electrocardiogram on Second Day of Life



Junctional bradycardia (heart rate 60 beats/min), with a prolonged corrected QT interval of 600 ms with a long ST-segment.

the setting of LA isomerism and ventriculoarterial concordance {A, D, S}. This rare condition is different from isolated atrial and ventricular inversion according to strict anatomic segmental descriptions but can be physiologically similar (4-8). An atrial switch operation such as the Senning procedure achieves

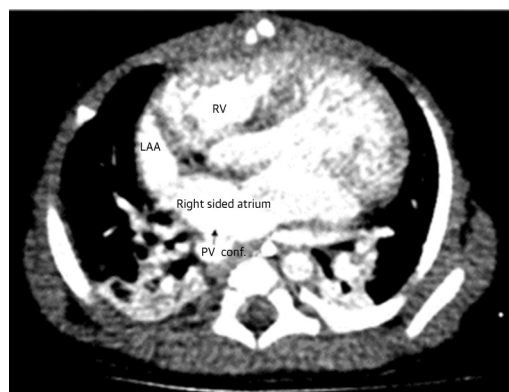
physiological correction by redirecting the pulmonary venous return into the systemic left ventricle and the systemic venous return into the subpulmonary right ventricle, as has been previously described (3-7).

FIGURE 4 Subcostal Short-Axis Echocardiographic Findings Showing the relationship of IVC, which is Anterior and Left to the AO

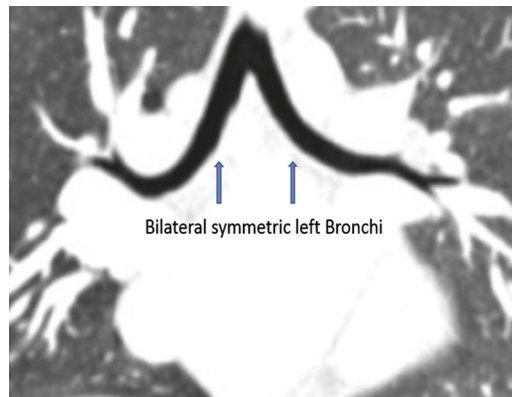


AO = aorta; IVC = inferior vena cava.

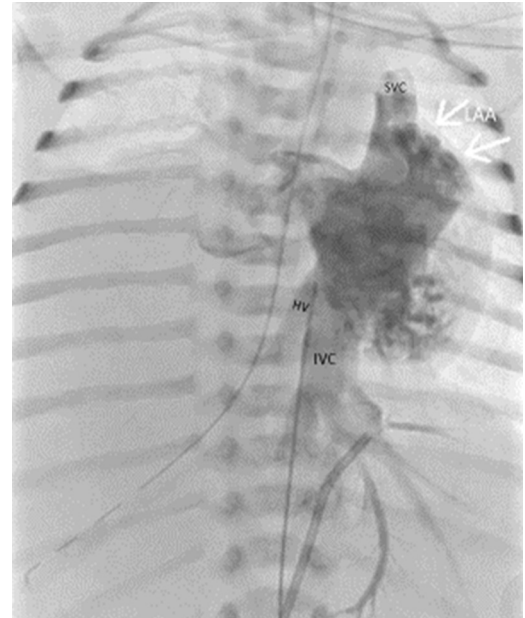
FIGURE 5 CTA Confirming Connection of Pulmonary Vein Confluence to a Right-Sided Atrium With a Morphological LAA



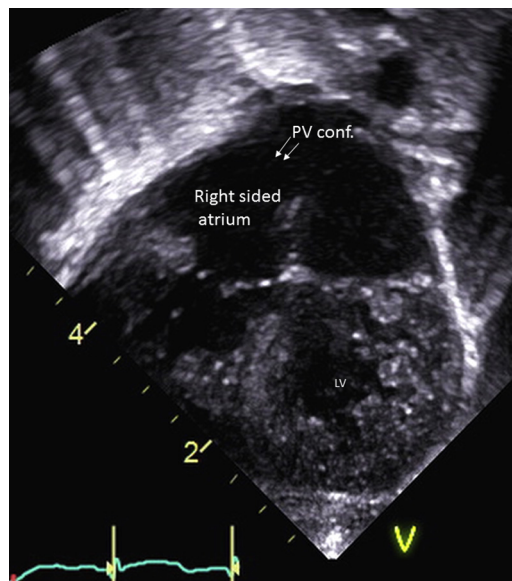
CTA = computed tomographic angiography; LAA = left atrial appendage; PV = pulmonary vein; RV = right ventricle.

FIGURE 6 CTA Confirming the Bilateral Morphological Left Bronchi

Junctional bradycardia due to a hypoplastic or absent sinus node, which is a right atrial structure, has also been well described in LA isomerism. In 30% of patients with LA isomerism, there are twin atrioventricular nodes, and pacing is required in 12% of patients (9). However, the association between long-QT

FIGURE 8 Atrial Angiogram Demonstrating the Connection of Systemic Veins to the Left-Sided Atrium With a Morphological LAA

HV = hepatic vein; IVC = inferior vena cava; LAA = left atrial appendage; SVC = superior vena cava.

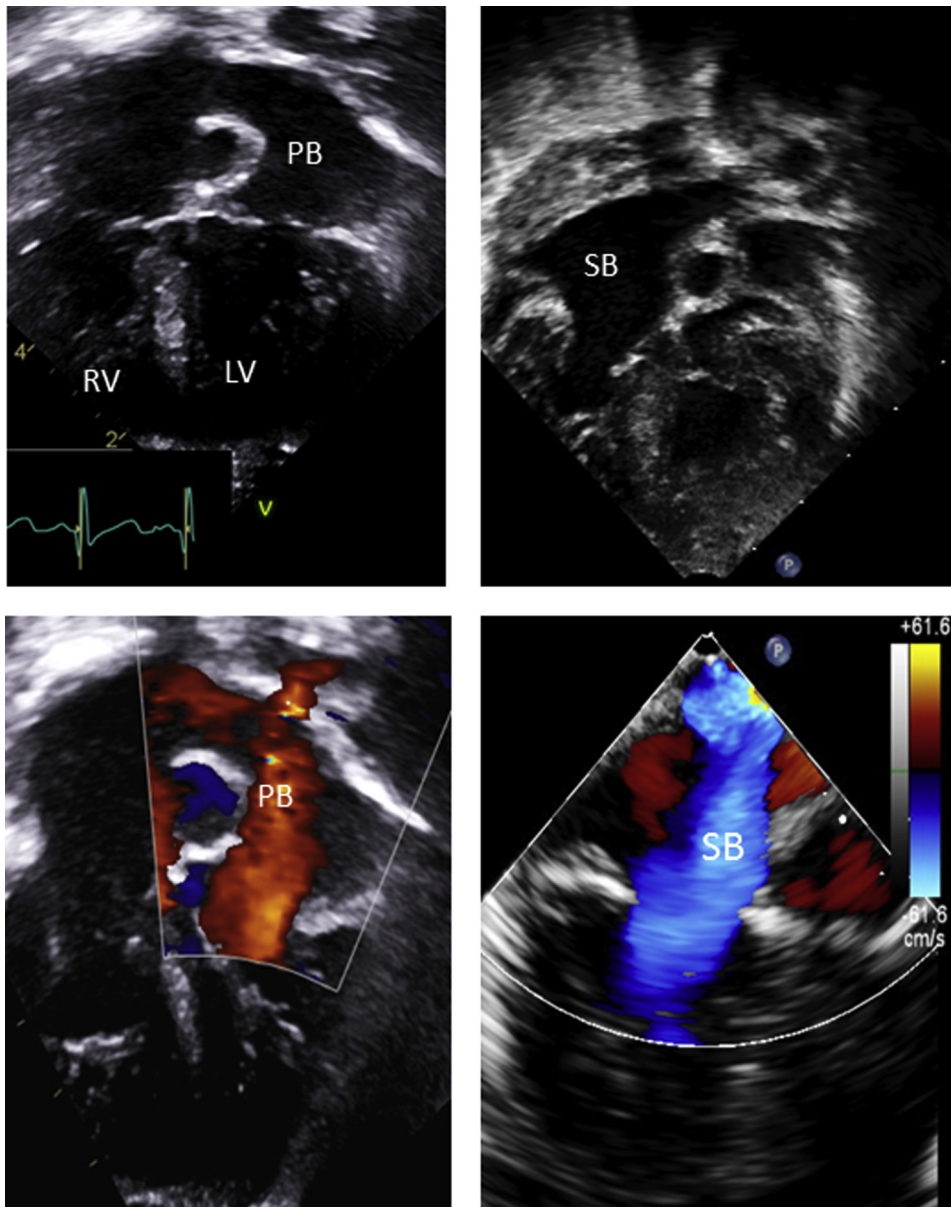
FIGURE 7 4-Chamber Echocardiographic View Showing Noncompaction of the Left Ventricle

Pulmonary venous (PV) confluence (conf.) is connected to the right-sided atrium.

syndrome and heterotaxy syndrome has not been described. Genetic testing for long-QT syndrome was negative in our patient, but he had an abnormal mutation in *DSC2* that is commonly associated with arrhythmogenic right ventricular cardiomyopathy. In long-term follow-up of atrial switch patients, significant atrial arrhythmias are described in about 20% (10), as are baffle obstruction or leakage, atrioventricular valve regurgitation, and ventricular dysfunction. Left ventricular noncompaction, noted in our patient, also has an increased prevalence among patients with heterotaxy syndrome (LA isomerism) compared with the general population (11).

Our patient remains at higher risk for chronic pulmonary disease and other related comorbidities not only secondary to CDH but also related to ciliary dysfunction associated with heterotaxy syndrome (12). Patients with LA isomerism exhibit a wide spectrum of anomalies involving the thoracoabdominal viscera, as was seen in our patient. On the basis of autopsy series of LA isomerism, intestinal malrotation is described in about 60.4% of patients (13). Our patient required Ladd's procedure because

FIGURE 9 Echocardiogram Demonstrating no Obstruction to Systemic or Pulmonary Intra-Atrial Baffle After Senning Procedure



LV = left ventricle; PB = pulmonary intra-atrial baffle; RV = right ventricle; SB = systemic intra-atrial baffle.

of the same, and a feeding gastrostomy tube was placed simultaneously. Urogenital anomalies have been reported in 6% of patients with LA isomerism (13), and the duplicated right kidney, hydronephrosis and nephrolithiasis, will continue to require long term follow up. The complexity of the diagnosis hence warrants a multi-disciplinary approach to care, to enhance outcomes and quality of life for these patients.

CONCLUSIONS

This exceedingly rare case demonstrates the complexity of multisystem anomalies that can present in a critically ill newborn. It also emphasizes a cardiac segmental nomenclature nightmare in complex congenital heart diseases and highlights the importance of meticulous, multimodal evaluation to accurately diagnose the condition and to optimize

management. Since the advent of the arterial switch operation for d-TGA, the role for an atrial switch operation such as a Senning or Mustard procedure has significantly diminished. However, as this case illustrates, there is still a role for the atrial switch operation outside of the more familiar diagnoses.

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REFERENCES

1. Senning A. Surgical correction of the transposition of the great arteries. *Surgery* 1959;45:966-80.
2. Jacobs JP, Andersen RH, Weinberg PM, et al. The nomenclature, definition and classification of cardiac structures in the setting of heterotaxy. *Cardiol Young* 2007;17 Suppl 2:1-28.
3. Uemura H, Ho SY, Devine WA, Kilpatrick LL, Anderson RH. Atrial appendages and venoatrial connections in hearts from patients with visceral heterotaxy. *Ann Thorac Surg* 1995;60:561-90.
4. Van Praagh R, Van Praagh S. Isolated ventricular inversion: a consideration of the morphogenesis, definition and diagnosis of not transposed and transposed great arteries. *Am J Cardiol* 1966;17:395-406.
5. McElhinney DB, Reddy VM, Silverman N, Hanley FL. Intraatrial baffle repair of isolated ventricular inversion with left isomerism. *Ann Thorac Surg* 1996;62:1529-32.
6. Sebastian VA, Guleserian KJ, Juraszek A, Kane C, Hamzeh R, Forbess JM. Modified Senning procedure for correction of atrioventricular discordance with total anomalous pulmonary venous return, atrial situs inversus, dextrocardia, and bilateral superior vena cavae. *Ann Thorac Surg* 2015;100:1446-8.
7. Santoro G, Masiello P, Farina R, Baldi C, Di Leo L, Di Benedetto G. Isolated atrial inversion in situs inversus: a rare anatomic arrangement. *Ann Thorac Surg* 1995;59:1019-21.
8. Clarkson PM, Brandt PW, Barratt-Boyes BG, Neutze JM. Isolated atrial inversion. Visceral situs solitus, viscerotrial discordance, discordant ventricular d-loop without transposition, dextrocardia: diagnosis and surgical correction. *Am J Cardiol* 1972;29:877-81.
9. Giljam T, McCrindle BW, Smallhorn JF, et al. Outcomes of left atrial isomerism over a 28-year period at a single institution. *J Am Coll Cardiol* 2000;36:908-16.
10. Jaillard SM, Pierrat V, Dubois A, et al. Outcomes at 2 years of infants with congenital diaphragmatic hernia: a population based study. *Ann Thorac Surg* 2003;75:250-6.
11. Martinez HR, Ware S, Schamberger S, Parent JJ. Noncompaction cardiomyopathy and heterotaxy syndrome. *Prog Ped Card* 2017;46:23-7.
12. Flinn CJ, Wolff GS, Dick M, et al. Cardiac rhythm after the Mustard operation for complete transposition of the great arteries. *N Engl J Med* 1984;310:1635-8.
13. Burton EC, Olson M, Rooper L. Defects in laterality with emphasis on heterotaxy syndromes with asplenia and polysplenia: an autopsy case series at a single institution. *Pediatr Dev Pathol* 2014;17:250-64.

KEY WORDS atrial inversion, left atrial isomerism, left ventricular noncompaction, Senning procedure