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Case Reports

Reviving the Atrial Switch: A Case Report of Successful Senning Procedure in a 13-Year-Old with Late-Presenting d-Transposition of the Great Arteries

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Abstract

Background: d-Transposition of the Great Arteries with an Intact Ventricular Septum (d-TGA-IVS) is a severe congenital cardiac anomaly typically necessitating an Arterial Switch Operation (ASO) during the neonatal stage. In late presenters with a regressed Left Ventricle (LV), ASO becomes high-risk or infeasible, making atrial switch procedures such as the Senning procedure a potential salvage option, particularly in resource-limited settings.

Case Illustration: We report a 13-year-old boy with lifelong cyanosis who developed progressive exertional dyspnea. Echocardiography and cardiac catheterization confirmed d-TGA-IVS with secundum atrial septal defect, severe Right Ventricular Hypertrophy (RVH), and preserved Pulmonary Vascular Resistance (PVR) (1.93 WU). Due to LV regression and the limited feasibility of late ASO, a Senning atrial switch was performed. Postoperative recovery was clinically stable, with oxygen saturation improving from 73% to 91%. Early postoperative echocardiography showed reduced systemic Right Ventricular (RV) function (Tricuspid Annular Plane Systolic Excursion [TAPSE] 7 mm), which subsequently improved on follow-up.

Conclusions: The Senning procedure remains a valuable salvage option for very-late presenting d-TGA-IVS with favorable pulmonary hemodynamics when ASO is not feasible. This case underscores the importance of periodic reassessment of operability in patients initially deemed inoperable and highlights the need for lifelong follow-up to monitor systemic RV function, arrhythmia risk, and baffle integrity.

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Keywords: d-Transposition of the great arteries, Senning procedure, late presenter, atrial switch, case report.

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Introduction

d-Transposition of the Great Arteries (d-TGA) represents roughly 5–7% of all congenital heart malformations and is the leading cause of cyanotic heart disease in newborns. Although prenatal screening has improved, a substantial number of cases, particularly in low- and middle-income countries, are only diagnosed postnatally, resulting in treatment delays.[1-2]

In cases of Intact Ventricular Septum (IVS), insufficient intercirculatory mixing causes marked hypoxemia. Absent timely surgical repair, the morphological Left Ventricle (LV) rapidly loses its conditioning after birth, diminishing its ability to support systemic circulation and making delayed arterial switch procedures more technically demanding and hazardous.[2]

Prior to the availability of the Arterial Switch Operation (ASO), the Senning and Mustard atrial switch techniques were the mainstay for d-TGA management, rerouting venous inflow within the atria to restore physiological flow. Although ASO provides better long-term outcomes, atrial switch procedures still have a role in carefully selected late-presenting or high-risk cases, especially in resource-limited settings.[3-4]

This case highlights an adolescent with late-presenting d-Transposition of the Great Arteries with an Intact Ventricular Septum (d-TGA-IVS) who underwent a successful Senning procedure, illustrating its ongoing relevance in carefully selected patients despite modern surgical trends.

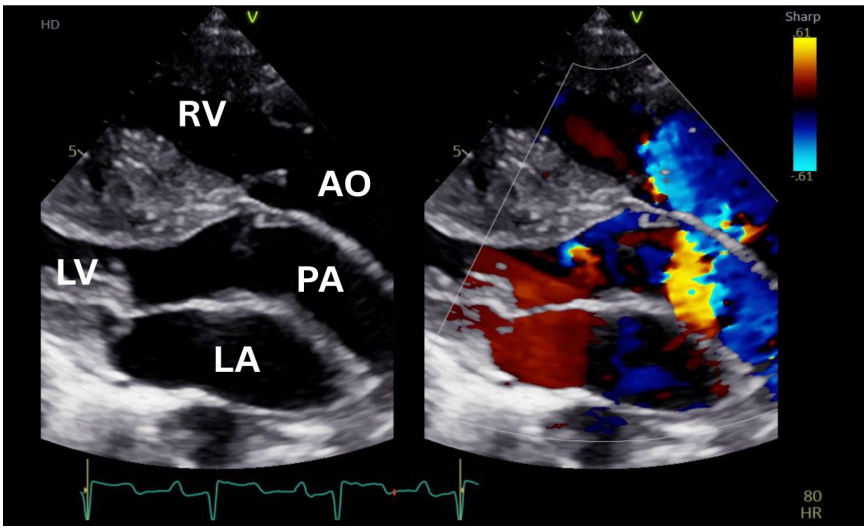

Figure 1. Preoperative transthoracic echocardiogram from the parasternal long-axis window, showing a markedly small, concentrically remodeled morphologic left ventricle (LV), reflecting left ventricular regression in this late-presenting d-TGA-IVS patient. The pulmonary artery is visualized, and color Doppler (right panel) confirms laminar antegrade flow in both pulmonary arteries.

Case Illustration

A 13-year-old boy was transferred from a secondary hospital with a confirmed diagnosis of d-TGA and a secundum atrial septal defect exhibiting a bidirectional shunt, accompanied by suspected pulmonary hypertension. The patient had been cyanotic since birth and had no history of prior cardiac surgery or interventions.

On initial transthoracic echocardiography, a parasternal long-axis view demonstrated situs solitus, concordant atrioventricular connections, discordant ventriculo-arterial alignment, an IVS, pronounced Right Ventricular Hypertrophy (RVH), moderate TR, and a markedly small, concentrically remodeled morphologic LV consistent with left ventricular regression (Figure 1). Pulmonary valve gradients were within normal limits. A more cranial parasternal long-axis sweep at the level of the pulmonary artery bifurcation showed the morphologic LV giving rise to the main pulmonary artery, which then divided into the right and left pulmonary arteries, confirming ventriculo-arterial discordance with a preserved pulmonary vascular bed (Figure 2).

On admission, oxygen saturation was 73%, blood pressure 91/63 mmHg, and heart rate 107

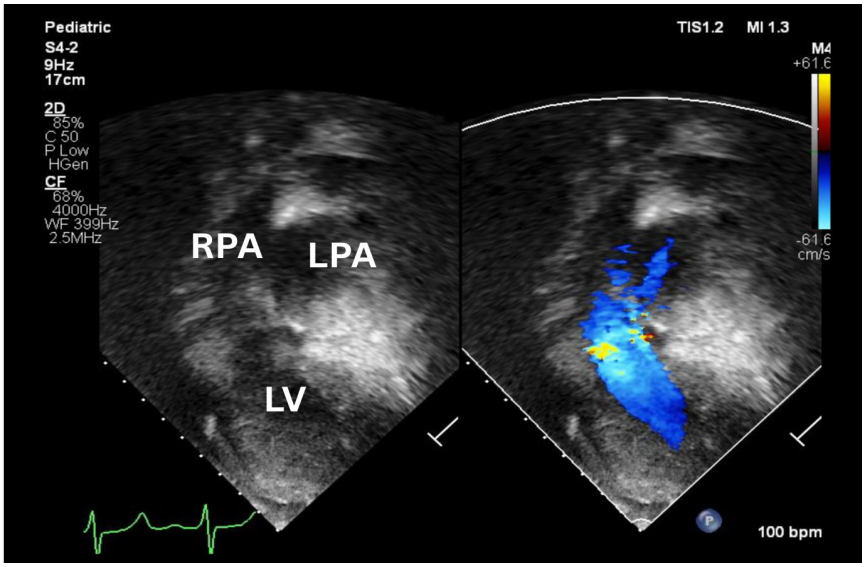

Figure 2. Preoperative transthoracic echocardiogram from the parasternal long-axis window at the level of the pulmonary artery bifurcation. The morphologic left ventricle (LV) gives rise to the main pulmonary artery, which then divides into the right (RPA) and left (LPA) pulmonary arteries. Color Doppler (right panel) shows laminar antegrade flow into both pulmonary artery branches, consistent with preserved pulmonary vascular bed and ventriculo-arterial discordance in d-TGA with an intact ventricular septum.

bpm. Physical examination revealed central cyanosis with clubbing, no audible murmurs, and clear lung fields. Laboratory results showed hemoglobin 16.5 g/dL and hematocrit 52.6%. Chest radiography demonstrated a normal cardiac silhouette without pulmonary congestion.

Cardiac catheterization confirmed systemic Right Ventricular (RV) pressures (82/7 mmHg), left ventricular pressures of 34/8 mmHg, and low Pulmonary Vascular Resistance (PVR) (PVR 1.93 WU; PVRi 1.91 WU·m^2; pulmonary vascular resistance/systemic vascular resistance ratio 0.16), indicating preserved pulmonary vascular reactivity. Coronary anatomy was unobstructed.

Due to marked left ventricular regression, elevated surgical risk, and the impracticality of performing a late ASO with ventricular retraining, the multidisciplinary team opted for an atrial switch (Senning) procedure. The operation involved rerouting pulmonary venous return to the morphologic right atrium and systemic venous return to the morphologic left atrium, with placement of two temporary RV pacing leads as standby. The patient remained hemodynamically stable after surgery. Early postoperative echocardiography (day 7) demonstrated unobstructed systemic and pulmonary venous pathways, good LV systolic function (EF 65%), and reduced systemic RV function with a Tricuspid Annular Plane Systolic Excursion (TAPSE) of 7 mm, likely reflecting early postoperative myocardial stunning and acute loading adaptation. A postoperative subcostal view (Figure 3) clearly showed the Senning pulmonary venous baffle, with pulmonary venous flow redirected to the morphologic right atrium without obstruction or leakage. Follow-up echocardiography at one month showed improvement in systemic RV systolic function, with TAPSE increasing to 13 mm. No significant pericardial effusion was detected. At the time of discharge, the patient's condition was stable, and oxygen saturation was 91% on ambient air.

Discussion

Introduced by Åke Senning in 1959, the Senning procedure was the primary surgical repair for d-TGA (d-TGA) until it was replaced by the ASO in the mid-1970s. The ASO became preferred because it re-establishes the LV, which is morphologically designed to support the systemic workload, thereby preventing the chronic complications associated with a systemic right ventricle in atrial switch repairs. Over subsequent decades, ASO has largely replaced atrial switch operations except in select high-risk or late-presenting patients whose LVs had regressed and could not sustain systemic pressures without retraining.[4-5]

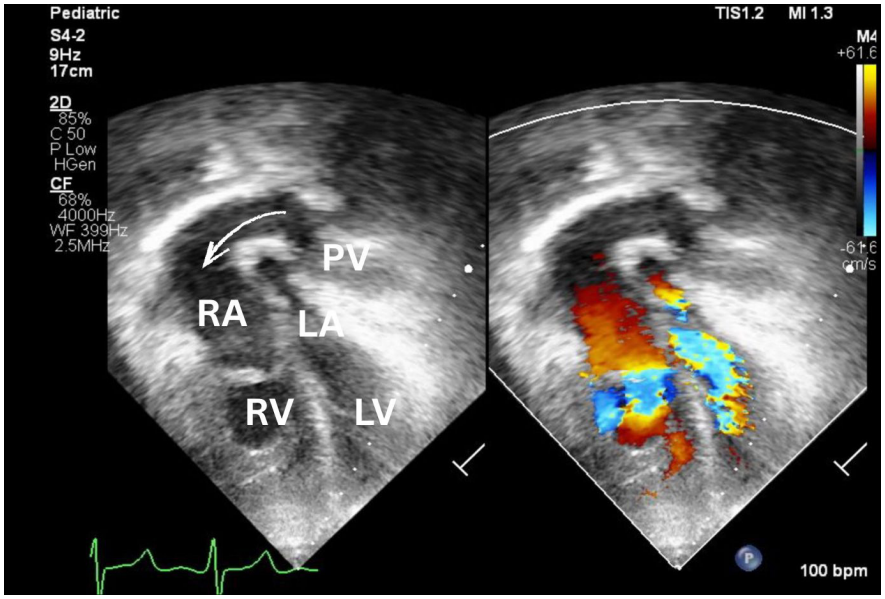

Figure 3. Postoperative subcostal transthoracic echocardiogram demonstrating the Senning pulmonary venous baffle (arrow). In the two-dimensional image (left panel), the pulmonary veins (PV) enter the baffle and are redirected toward the morphologic right atrium (RA), with the morphologic left atrium (LA), right ventricle (RV), and left ventricle (LV) also visualized. Color Doppler imaging (right panel) shows laminar flow through the pulmonary venous baffle into the RA without obstruction or leakage, consistent with a functioning atrial switch repair.

Late clinical presentation of d-TGA accompanied by an IVS is uncommon. If left untreated, the condition results in death in as many as 90% of patients in their first year of life, with only a minimal proportion living into adolescence or adulthood without operative intervention. Our patient's survival to 13 years with preserved exercise tolerance despite chronic cyanosis is therefore highly unusual. Similar to the few reports describing survival into the third and fourth decades, our patient demonstrated unexpectedly preserved functional capacity despite longstanding cyanosis.[5-6] We observed that preserved PVR (PVR 1.93 WU) allowed the surgical team to safely proceed with the Senning procedure without posing a prohibitive risk of systemic RV failure or fixed pulmonary hypertension.[5-6]

This finding reinforces the importance of periodic reassessment of operability in patients initially deemed inoperable, as pulmonary vascular adaptation can vary widely among individuals and may be influenced by interatrial shunt size, early pulmonary artery pressures, and possibly genetic factors.[7]

Alternative surgical strategies for such late presenters, such as late ASO with left ventricular retraining or heart transplantation, are often limited by feasibility, cost, and availability in resource-limited settings. In this context, the Senning procedure remains a valuable option, offering physiological correction and meaningful improvements in oxygen saturation and functional status, albeit with the trade-off of potential long-term complications, including systemic RV dysfunction, atrial arrhythmias, and baffle obstruction.[8–10]

Despite the availability of a more preferred surgical procedure, the Senning atrial switch procedure may still provide optimal outcomes, particularly in late-presenting patients. A similar case report has chosen the Senning method instead of ASO in consideration of the patient's acceptable hemodynamic parameters.[11] Additionally, this method is said to provide a 'simple and effective' modality, particularly for patients with a late presentation. Another case report unanimously agreed that atrial switch is the ideal option for a late-presenting patient, which is similar to the rationale used for the patient in this case report.[12] In late-presenting patients, primary ASO may be less ideal, as previously documented literature has only reported the success of LV retraining without prior atrial switch.[13] Therefore, this procedure may be opted for in cases that do not fit the prerequisites of an ASO.

Conclusion

The Senning atrial switch continues to serve as a life-saving surgical option in adolescents with very late-presenting d-TGA-IVS, particularly when ASO is unfeasible due to left ventricular regression or resource limitations. Our patient's favorable outcome, with improved oxygenation and stable hemodynamics, reinforces the importance of individualized surgical decision-making and periodic reassessment of operability. Ongoing long-term surveillance is crucial for identifying systemic RV dysfunction, atrial arrhythmias, and complications involving the baffle.

List of Abbreviations

ASO	Arterial Switch Operation
d-TGA	d-Transposition of the Great Arteries
d-TGA-IVS	d-Transposition of the Great Arteries with an Intact Ventricular Septum
IVS	Intact Ventricular Septum
PVR	Pulmonary Vascular Resistance
RV	Right Ventricular
RVH	Right Ventricular Hypertrophy
TAPSE	Tricuspid Annular Plane Systolic Excursion

Ethical Clearance

Not applicable.

Publication Approval

Obtained.

Authors Contributions

R.M., S.A., and S.N.S conceived the study idea and concept. R.M., S.A., I.K.M, A.R, E.O.J., D.F., R.S.A, H.A., and M.A.A contributed to the study design and preparation of figures. S.N.S and M.A.A supervised the project. RM., H.A., S.N.S., and M.A.A drafted the manuscript. All authors reviewed, commented, and approved the final version of the manuscript.

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Conflict of Interest

None.

Availability of Data and Materials

Not applicable.

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Not applicable.

Generative AI and AI-Assisted Technologies in the Writing Process

Authors acknowledge that artificial intelligence (AI) tools were only used to assist in language editing and did not generate or alter the scientific content, analyses, or conclusions presented in this manuscript.

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