Dextro-transposition of the great arteries coexisting with severe non-diabetic hypertrophic cardiomyopathy in a late-presenting infant: a sentinel case and review of surgical limitations

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ABSTRACT

Background: Co-occurrence of dextro-transposition of the great arteries (D-TGA) and severe hypertrophic cardiomyopathy (HCM) is an exceptionally rare and hemodynamically precarious scenario. While transient, reversible HCM is well-documented in neonates of diabetic mothers, idiopathic biventricular HCM presenting late in infancy introduces unique physiological barriers.

Case Presentation: We present the case of a 4-month-old, non-diabetic infant diagnosed with parallel circulation D-TGA, complicated by massive biventricular hypertrophy, a large ventricular septal defect, and flow-related pulmonary hypertension.

Discussion: The contemporaneous presentation of these pathologies creates a vicious hemodynamic cycle: the excessive volume and pressure loading from unrestrictive shunts collide with severe intrinsic biventricular hypertrophy, culminating in profound diastolic dysfunction and severely restricted inter-circulatory mixing. This case underscores that, unlike transient neonatal HCM, late-presenting non-diabetic hypertrophy combined with high-volume pulmonary overcirculation renders traditional bridging and corrective therapies physiologically intolerable.

Conclusion: We detail the profound surgical limitations in this subset of patients - specifically the theoretical contraindications for pulmonary artery banding, the Arterial Switch Operation, and atrial switch procedures. We highlight why formal surgical atrial septectomy, with or without patent ductus arteriosus ligation, is proposed as a potential salvage pathway to navigate such profound physiological fragility when advanced percutaneous interventions are unavailable.

Keywords: D-transposition of the great arteries, congenital anomalies, hypertrophic cardiomyopathy.

Type of Article: CASE REPORT **Specialty:** Cardiology

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Background

Congenital heart disease affects approximately 1% of live births globally [1], with dextro-transposition of the great arteries (D-TGA) representing a critical cyanotic anomaly prevalent in 2 to 5 per 10,000 live births [2]. Survival in D-TGA is strictly contingent upon adequate inter-circulatory mixing. When D-TGA is compounded by hypertrophic cardiomyopathy (HCM), the clinical paradigm shifts dramatically. Historically, this dual pathology is most frequently observed in infants of diabetic mothers (IDM), where hyperinsulinemia drives transient myocardial thickening that typically regresses postnatally [3]. However, the presence of severe, biventricular HCM in a non-diabetic, late-presenting infant is exceedingly rare and poorly represented in the literature. This case highlights the devastating compounding effect of extreme, fixed hypertrophy and flow-related pulmonary hypertension on parallel circulation physiology, radically challenging the efficacy of established palliative and surgical timelines.

Case Presentation

A 4-month-old female infant (weight: 5.4 kg) was referred for urgent cardiac evaluation due to profound cyanosis, respiratory distress, and differential hypoxemia (right upper limb SpO_2: 40%; right lower limb SpO_2: 64%). The maternal history was uncomplicated; a 75g oral glucose tolerance test at 26 weeks of gestation was negative for gestational diabetes. A detailed three-generation family history revealed no instances of cardiomyopathy, unexplained infant death, or sudden cardiac death. Physical examination lacked dysmorphic features suggestive of Noonan syndrome or other RASopathies.

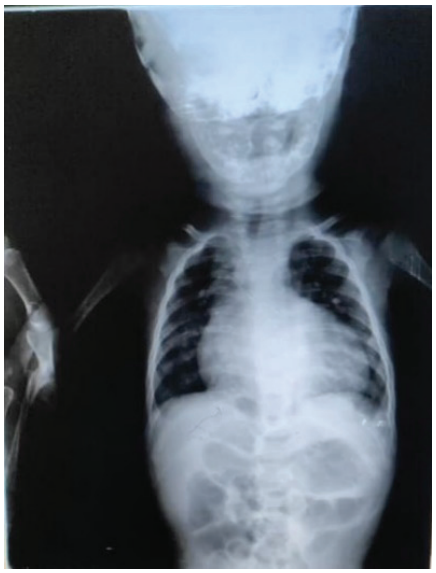

Figure 1. *Chest radiography revealed increased pulmonary vascular markings and the classic "egg-on-a-string" cardiac silhouette.*

Initial chest radiography revealed increased pulmonary vascular markings and the classic "egg-on-a-string" cardiac silhouette (Figure 1). Transthoracic echocardiography demonstrated situs solitus, atrioventricular concordance, and ventriculo-arterial discordance consistent with parallel circulation D-TGA. Notably, the right ventricle was severely hypertrophic, with extreme thickening of the anterior wall (RVAW: 10 mm; normal reference for age and weight typically < 3 mm). The left ventricle exhibited profound concentric hypertrophy, evidenced by extreme thickening of both the interventricular septum (IVSd: 11 mm, Z-score: +4.8) and the posterior wall (LVPWd: 10 mm, Z-score: +5.8). This culminated in a severely elevated left ventricular mass index (LVMI: 199 g/m²), as calculated by echocardiography.

Further echocardiographic assessment identified a tricuspid pulmonary valve with marked dilation of the main pulmonary trunk and its respective branches. Doppler

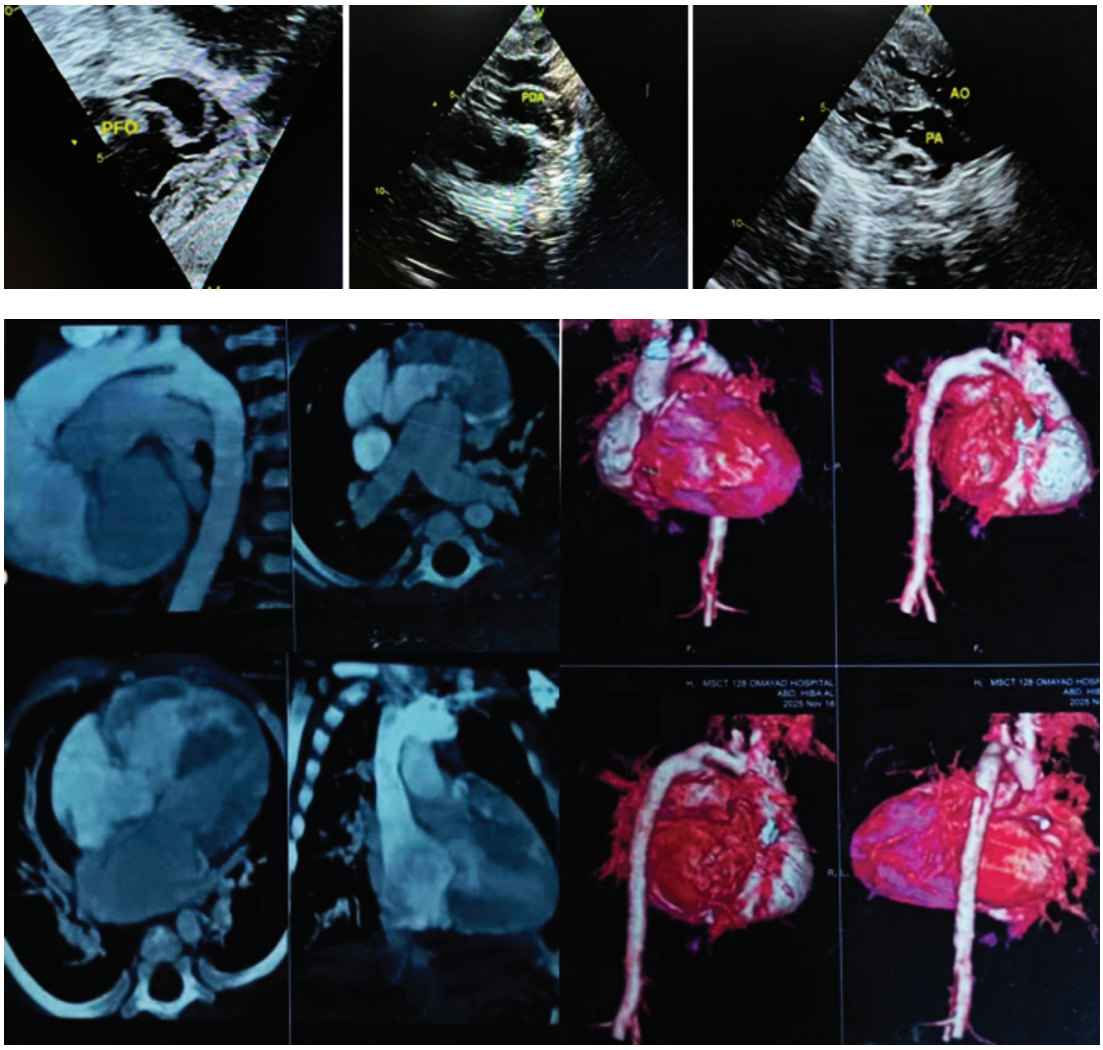

Figure 2*. Transthoracic echocardiography demonstrated situs solitus, atrioventricular concordance, and ventriculo-arterial discordance consistent with parallel circulation D-TGA. Notably, the right ventricle was severely hypertrophic, with extreme thickening of the anterior wall, a large patent ductus arteriosus (4 × 5 mm), an inlet ventricular septal defect (6 × 9 mm), and a patent foramen ovale (4 × 5 mm).*

interrogation demonstrated a peak transvalvular velocity of 1.2 m/s - accompanied by mid-systolic closure, a hallmark echocardiographic sign of pulmonary hypertension driven by excessive pulmonary blood flow. Inter-circulatory mixing was facilitated by a triad of bidirectional shunts: a large patent ductus arteriosus (PDA), an inlet ventricular septal defect (VSD), and a patent foramen ovale (PFO). Despite the massive structural biventricular hypertrophy, the left ventricle maintained normal chamber dimensions. This preserved cavity size is a direct physiological consequence of the concurrent volume loading from the unrestrictive shunts counterbalancing the pressure-induced concentric inward remodeling (Figure 2; Table 1).

A multi-slice computed tomography (MSCT) angiogram provided definitive spatial corroboration of the complex conotruncal anatomy. The anterior ascending aorta (14 mm) originated from the morphologic right ventricle with a left-sided aortic arch (6 mm) and normal coronary origins. The posterior pulmonary trunk (19 mm) originated from the morphologic left ventricle. No aortic coarctation was present (Figure 3).

Table 1. *Brief description of the case.*

Parameter	Measurement/observation	Reference/normal value
Right ventricular anterior wall (RVAW)	10 mm	< 3 mm
Interventricular septum (IVSd)	11 mm (Z-score: +4.8)	Normal limits for age/weight
Left ventricular posterior wall (LVPWd)	10 mm (Z-score: +5.8)	Normal limits for age/weight
Left ventricular mass index (LVMI)	199 g/m²	< 65 g/m²
Ventricular function	Preserved LV cavity size, impaired relaxation	Normal biventricular compliance
Main pulmonary artery (MPA)	18 mm (Dilated)	-
Estimated pulmonary pressure	Peak velocity 1.2 m/s with mid-systolic closure	Suggestive of severe PAH
Shunt directions	PDA (4 × 5 mm), VSD (6 × 9 mm)	Bidirectional
Atrial communication	PFO (4 × 5 mm)	Restrictive flow

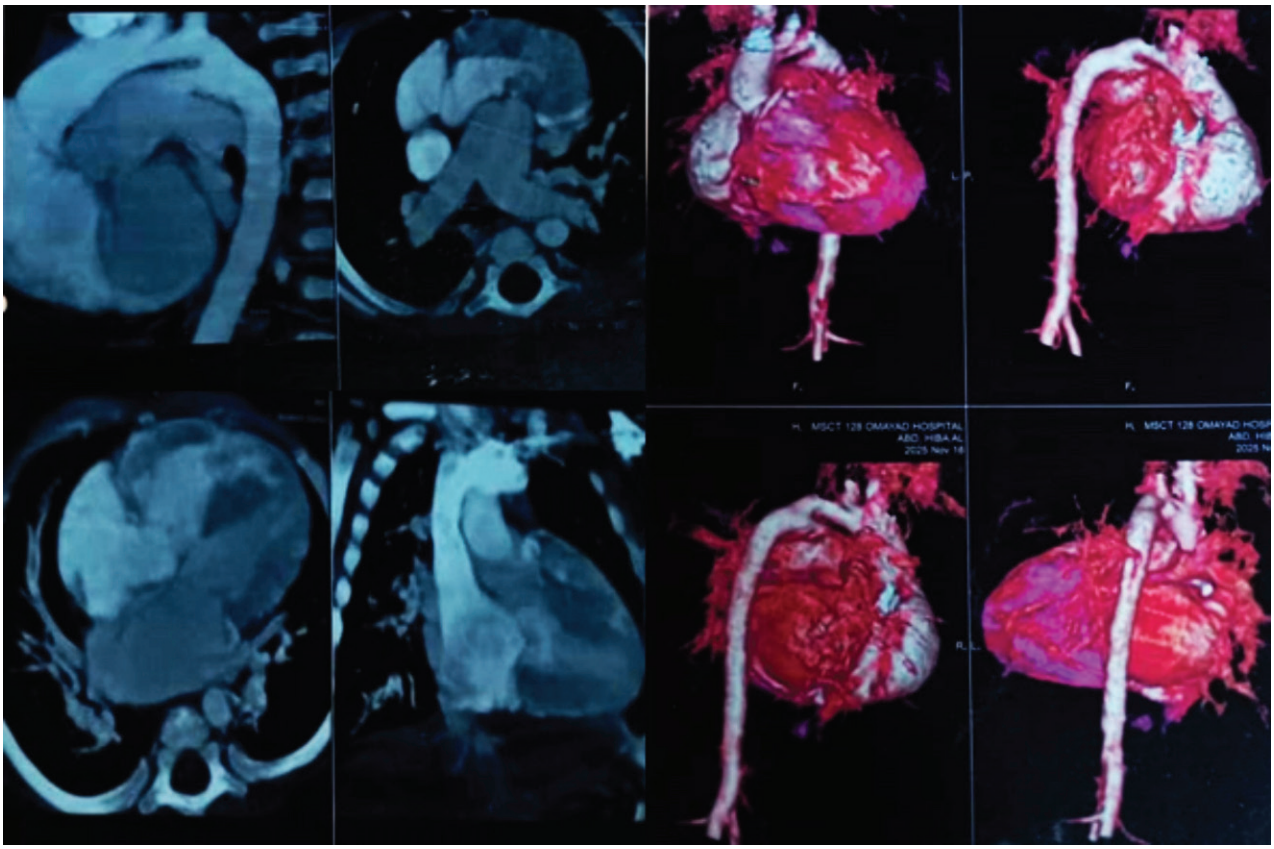

Figure 3*. A MSCT angiogram provided definitive spatial corroboration of the complex conotruncal anatomy.*

Table 2. *Comprehensive serum analyses.*

ANALYTE	MEASURED VALUE	REFERENCE RANGES
Lactate	44 mg/dl	8.1-15.3 mg/dl
Glucose	85 mg/dl	70-110 mg/dl
Ammonia	166 µg/dl	<245 µg/dl
Pyruvate	3.47 mg/dl	0.7-1.4 mg/dl
TSH	1.6 IU/ml	0.27-4.20 IU/ml

Extensive genetic and metabolic workups were conducted to elucidate the underlying etiology of the myocardial hypertrophy. Both an in-house Exome Xtra enrichment panel and whole-exome sequencing yielded negative results, identifying neither pathogenic copy number variants nor underlying metabolic defects. Subsequent metabolic screening specifically ruled out glycogen storage diseases, such as Pompe disease, and comprehensive serum analyses confidently ruled out an acute metabolic crisis (Table 2).

Following a comprehensive multidisciplinary review, standard definitive and palliative surgical options were deemed exceptionally high risk due to the severe, non-compliant nature of the biventricular hypertrophy. The rapid progression of the clinical course is outlined below:

Clinical timeline

- Day 1 (admission and initial imaging): Patient admitted with profound cyanosis (SpO_2 40-64%). Initial echocardiography identified parallel circulation D-TGA with severe biventricular HCM. Prostaglandin E1 infusion was initiated to maintain ductal patency.
- Day 2 (advanced imaging and workup): MSCT angiogram confirmed conotruncal anatomy and excluded aortic coarctation. Extensive metabolic, syndromic, and genetic panels were sent.
- Day 3 (multidisciplinary team decision): The multidisciplinary team concluded standard corrective options (ASO, Senning) were physiologically unsuitable. Urgent surgical atrial septectomy was agreed upon as the required salvage pathway, driven by the absence of specialized percutaneous equipment.
- Day 4 (hemodynamic collapse): The patient experienced rapid, progressive diastolic dysfunction and refractory hypoxemia, leading to cardiac arrest. Unfortunately, resuscitation attempts failed, and the patient died before the scheduled surgical intervention could be performed.

Discussion

D-TGA is a life-threatening conotruncal anomaly where early mortality is averted only by adequate mixing of the parallel circulations [2,4]. In our patient, baseline hemodynamics were catastrophically complicated by extreme, intrinsic biventricular thickening [5]. Crucially, the sheer magnitude of this myocardial hypertrophy (LVMI: 199 g/m², IVS Z-score: +4.8) profoundly exceeds any expected adaptive remodeling induced by D-TGA, an unrestrictive VSD, or pulmonary hypertension. This extreme disproportionality definitively isolates a severe HCM phenotype as the cornerstone of this case [6].

While our ultimate consensus prioritized an urgent surgical septectomy to guarantee obligate inter-circulatory mixing, the physiological intent of this maneuver diverged strictly from historical paradigms. Previous literature frequently describes the successful management of D-TGA with concurrent hypertrophy using a delayed surgical approach [3], utilizing temporary interventions such as balloon atrial septostomy (BAS) to afford the myocardium time to naturally regress. However, that expectation of spontaneous regression is exclusively validated for transient, metabolically driven secondary hypertrophy - such as that observed in IDM. Extrapolating this delayed, expectant strategy to a late-presenting, non-IDM infant afflicted with fixed biventricular hypertrophy constitutes a profound physiological miscalculation.

The presence of this primary, non-regressive HCM abruptly closes traditional therapeutic windows, systematically eliminating every standard surgical avenue:

- Arterial switch operation (ASO): Definitive anatomical repair via the ASO is rendered physiologically unviable. While the ASO requires the sub-pulmonary left ventricle to safely assume systemic vascular resistance, this patient's LV is already intrinsically diseased and severely hypertrophied. Subjecting such pathologically non-compliant myocardium to the obligate stressors of cardiopulmonary bypass, ischemic arrest, and coronary transfer carries a prohibitive risk of acute myocardial failure and low cardiac output syndrome.
- Atrial switch procedures: Physiological correction utilizing an atrial switch (Senning or Mustard procedure) is precluded by the patient's profound diastolic dysfunction. Because both chambers are pathologically stiff, forced venous rerouting would generate prohibitively high end-diastolic pressures, inevitably precipitating simultaneous and catastrophic pulmonary and systemic venous congestion.
- Pulmonary artery banding (PAB): Imposing an acute, fixed pressure afterload on an already intrinsically hypertrophic ventricle exacerbates wall stress and dramatically increases myocardial oxygen demand. Rather than providing a bridge to stability, this maneuver worsens subendocardial ischemia and invariably triggers acute diastolic and systolic collapse.

It is critical to acknowledge that the contraindications detailed regarding the ASO, atrial switch, and PAB in this specific patient are grounded in physiological reasoning, echocardiographic parameters, and extrapolation from

known hemodynamic principles, rather than direct empirical evidence from failed surgical attempts.

Furthermore, while standard BAS is often utilized for initial palliation, it is inherently insufficient in this late-presenting phenotype [7]. By 4 months of age, the naturally thickened and muscular atrial septum resists traditional balloon avulsion, frequently leaving a restrictive tissue flap against stiff, non-compliant ventricles. A procedure capable of excising tissue - such as a percutaneous blade septostomy - is optimal. However, due to the lack of requisite catheterization equipment for blade septostomy at our center, our institutional protocol dictates a pivot to urgent formal surgical atrial septectomy. This surgical approach ensures the permanent, low-resistance mixing chamber necessitated by the equipment constraints of our clinical environment. Furthermore, the proposed addition of PDA ligation offered a targeted mechanism to deliberately alleviate volume-driven pulmonary overcirculation.

Conclusion

While the contemporaneous presentation of D-TGA and primary, fixed biventricular hypertrophy is exceptionally rare, the rationale for our tailored approach is firmly anchored in absolute hemodynamic constraints. In the presence of profound, non-reversible hypertrophy, standard corrective timelines (arterial or atrial switch operations) and conventional surgical palliations (such as pulmonary artery banding) are rendered physiologically obsolete and highly lethal. Furthermore, transcatheter BAS is entirely unsuitable in this late-presenting phenotype.

Ultimately, due to anatomical barriers and institutional equipment constraints precluding blade septostomy, the planning of an urgent formal surgical atrial septectomy was necessitated. By definitively excising the obstructing tissue, this approach aims to guarantee the permanent, unrestrictive communication mandated by the severely reduced ventricular compliance. Combined with PDA ligation to mitigate volume-driven pulmonary overcirculation, this surgical strategy is proposed as a potential salvage pathway, theoretically prioritizing obligate inter-circulatory mixing and targeted hemodynamic stabilization in a profoundly fragile state.

What's new?

The presence of severe biventricular HCM in a non-diabetic is exceedingly rare and poorly represented in the medical literature.

List of Abbreviations

ASO	Arterial Switch Operation
BAS	Balloon atrial septostomy
D-TGA	Dextro-transposition of the great arteries
HCM	Hypertrophic cardiomyopathy
IDM	Infants of diabetic mothers IDM
OGTT	Oral glucose tolerance test
PAB	Pulmonary artery banding

Ethical approval

Ethical approval for this study was obtained. A copy of the ethical approval is available for review by the Editor-in-Chief of this journal.

Conflicts of interest

The authors declare that they have no conflict of interest regarding the publication of this case report.

Data availability

We declare that there is an electronic copy of patient data available for review by the Editor-in-Chief of this journal.

Consent to participate

A consent to participate in the advertisement in the manuscript was obtained from patients' parents. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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Summary of case

1	Patient (gender, age)	Female/4-month-old
2	Final diagnosis	D-TGA is compounded by Hypertrophic Cardiomyopathy (HCM)
3	Symptoms	Cyanosis, respiratory distress, and differential hypoxemia
4	Medications	None
5	Clinical procedure	Atrial septectomy
6	Specialty	Pediatric cardiology