CASE STUDY

106. Surviving The Odds: A Rare Case of Untreated Transposition Of The Great Arteries In Adulthood With Neurological Manifestations

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Background: Dextro- transposition of the great arteries (dTGA) is a very rare congenital cardiac anomaly with an incidence of 0.02-0.05% that usually results in 90% infantile mortality, without surgical correction. Simultaneous interposition with other anomalies like atrial septal defect (ASD), ventricular septal defect (VSD), can enable survival beyond infancy, in some instances. However, long-term survival into adulthood remains exceptionally uncommon, especially when complicated with brain abscess- a known but serious complication of cyanotic CHD.

The Case: We present a case of Cyanotic congenital heart disease with d-TGA, VSD, ASD, dextro-cardia, complicated by a cerebral abscess.

A 28-year-old male presented with two episodes of focal seizures and one episode of loss of consciousness. The patient was previously diagnosed with cyanotic congenital heart disease at age 7 months, but chose not to opt for surgical intervention as there was no delay in developmental & intellectual milestones. Since then, he had multiple episodes of deepening of cyanosis with palpitations & giddiness, which were subsequently managed by blood letting & incomplete staged surgery.

Upon arrival at our hospital, he developed weakness over left-half of his body, which resolved spontaneously. On examination he was conscious and oriented. He also had, prominent bilateral EJV with central cyanosis and Grade 2 clubbing. CVS examination revealed Grade III mid-systolic murmur all over the precordium and apex beat at right sixth intercostal space along mid-clavicular line. CNS examination revealed no focal neurological deficit. All other systems were unremarkable.

Echocardiography and cardiac catheterization confirmed:

- Situs solitus with dextrocardia
- Large ASD and VSD with common atrium
- Pulmonary artery & aorta arising parallelly (dTGA)
- Severe valvular and infundibular pulmonary stenosis (PS)
- Hypoplastic right ventricle and normal left ventricular function

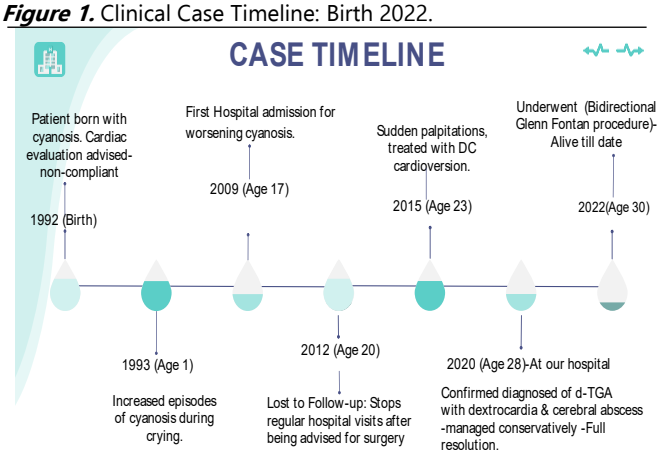
MRI brain revealed a right parietal lobe cerebral abscess. Blood and urine cultures were sterile.

The patient was deemed unfit for neurosurgical drainage due to high cardiac risk. He was treated with IV vancomycin, ceftriaxone, and metronidazole for 6 weeks. He showed significant clinical improvement and was discharged hemodynamically stable. Two years later, he successfully underwent Bidirectional Glenn → Fontan procedure with PA banding and atrial septectomy.

The patient remains alive & clinically stable till date and continues to come for periodic cardiology follow-up and has had no recurrence of cardiovascular or neurological complications.

Conclusion: In the present case, the VSD and atrial septal defect served as effective admixture lesions, permitting intermixing of systemic and pulmonary circulation, thereby maintaining some degree of oxygen saturation. Severe pulmonary limited pulmonary overcirculation, protecting the pulmonary vasculature and enabled long-term survival. However, long-standing right-to-left shunting predisposes to cerebral abscess and stroke, due to paradoxical embolization and impaired immune function. This case underscores the interplay of admixture lesions (VSD, ASD), protective pulmonary stenosis, leading to survival into adulthood in unoperated d-TGA.

Figure 1. Clinical Case Timeline: Birth 2022.



ISSN 2076-6327

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