

## Pulmonary endarteritis, cerebral abscesses, and a single ventricle: An uncommon combination

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### ABSTRACT

Endocarditis of the right side of the heart is otherwise uncommon in children. Pulmonary endarteritis as a complication of congenital heart disease is even rarer. Herein, we report the case of pulmonary endarteritis with a 7 mm ×5 mm vegetation, involving the main pulmonary artery in a 4-year-old male child, with cyanosis and a 1-week history of fever and rapidly-progressive hemiparesis. A full segmental echocardiography demonstrated a double inlet left ventricle with left-sided subaortic hypoplastic right ventricle (Van Praagh's A-III type – Single Ventricle). Additionally, CT scan of the brain revealed bilateral cerebral abscesses. To the best of our knowledge, the occurrence of pulmonary endarteritis and cerebral abscesses in a case of single ventricle is hitherto unreported. This article underlines the importance of heightened clinical awareness and meticulous echocardiography in cases of congenital heart disease so that relatively rare complications may not be missed.

**Key words:** Cerebral abscess, cyanotic heart disease, pulmonary endarteritis, single ventricle

### INTRODUCTION

Endocarditis of the right side of the heart is uncommon by virtue of the low hemodynamic pressure and lack of isolated or significant right-sided valvular deformities. The valves fall prey to an infective process in settings of congenital heart disease or endothelial injury.<sup>[1]</sup> Pulmonary arterial endarteritis is a rare event, even in patients with congenital heart disease (CHD).<sup>[2]</sup> Further, a combination of pulmonary endarteritis with a systemic suppuration in CHD invites greater attention.

### CASE REPORT

A 4-year-old male child, born from a non-consanguineous

marriage, was admitted with the history of fever and progressive right-sided weakness for 1 week and cyanosis noted since 6 months of age. The patient's mother had noted dyspnea on feeding since 1 year of age. On admission, the patient had dyspnea on exertion (grade III/IV, ATS grading). There was no history of cyanotic spells. The child was delivered vaginally at home and was unsupervised. The delivery process was uneventful. The patient started walking at 18 months, and motor milestones were delayed.

The child weighed 10 Kg., was febrile, irritable, tachycardic, and dyspneic. Significant cyanotic burden and clubbing were present. Chest was clear and vesicular breath sounds audible. The cardiac apex was at left 5<sup>th</sup> intercostal space, 3 cm outside the mid-clavicular line. An ejection systolic murmur (grade III/VI) was audible in the pulmonary area. A right-sided complete hemiparesis with extensor plantar was noted.

Investigations revealed a polymorphonuclear leukocytosis, hemoglobin - 17 gm% and PCV – 60%. An ECG revealed right atrial enlargement, extreme

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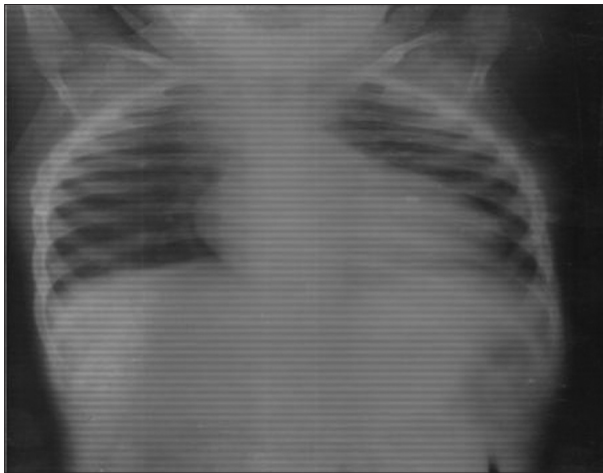
right axis, and poor progression of R wave. Chest x-ray revealed cardiomegaly with reduced pulmonary vascular markings [Figure 1]. A full-segmental echocardiography revealed a single ventricle of the double-inlet left ventricle type [Figure 2]. A rudimentary right ventricle was connected to the main ventricle by a non-restrictive foramen. The great vessels were malposed – the aorta arising from the rudimentary chamber and pulmonary artery from the main chamber [Figure 3]. Pulmonary stenosis with peak systolic gradient of 40 mmHg was seen. Pulmonary arteries were hypoplastic. A 7 mm × 5 mm fixed structure on the wall of the main pulmonary artery with erratic movement, indicative of a vegetation, suggested pulmonary endarteritis [Figure 4a and b]. A small ostium-secundum atrial septal defect was present, but patent ductus arteriosus was not present. CT scan of the brain revealed

bilateral multiple cerebral abscesses [Figure 5], with the largest one present superficially in the left parietal region. Blood culture showed growth of streptococcus pyogenes.

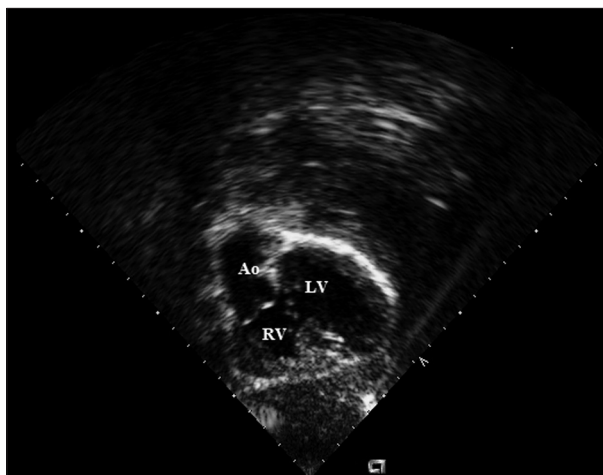
The child was started on parenteral ceftriaxone – 80 mg/ Kg/day (plus, gentamicin – 3 mg/Kg/day for 2 weeks) for the endarteritis and was continued for 6 weeks for optimal treatment of the cerebral abscess. Dyspnea improved with low dose intravenous frusemide. An attempted drainage of the largest cerebral abscess resulted in some improvement of the hemiparesis. The patient became afebrile in 2 weeks and was able to walk after 4 weeks of therapy. Repeat echo before discharge showed substantive decrease in size of the pulmonary artery vegetation and cardiac symptoms. The patient has been referred to the cardiac surgery department for corrective surgery for single ventricle.

## DISCUSSION

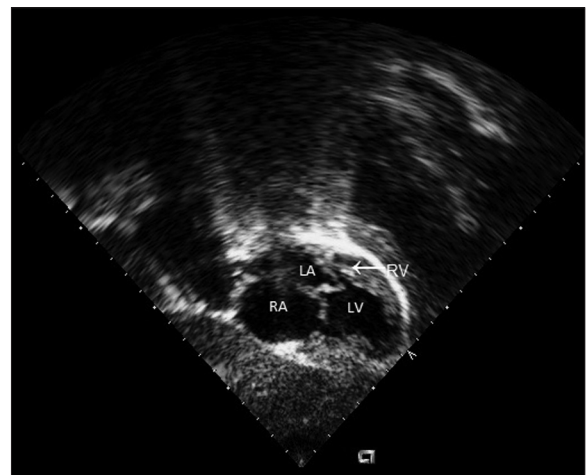
The risk of infective endocarditis is considerably increased in children with congenital heart disease, and the disease can occur with almost any lesion, more



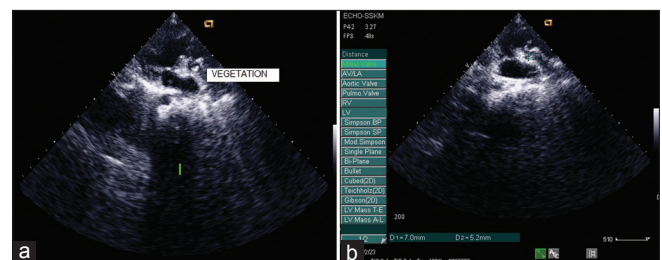
**Figure 1:** Chest X-ray showing cardiomegaly with decreased pulmonary flow



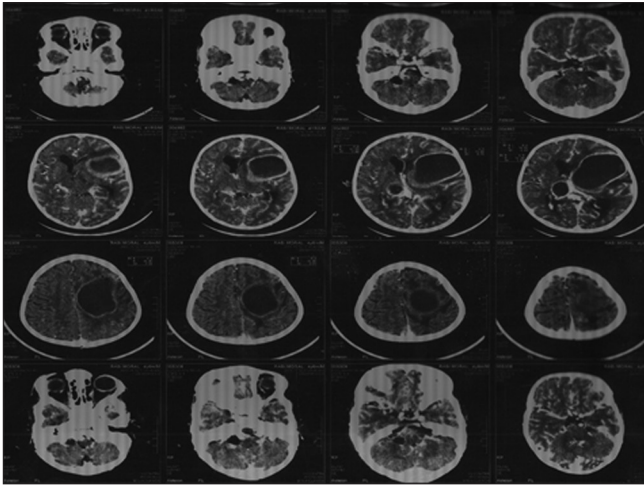
**Figure 3:** Echocardiography (subcostal anatomically corrected view) – showing aorta arising from a rudimentary right ventricle which is connected to the left ventricle through a nonrestrictive foramen



**Figure 2:** Echocardiography (subcostal anatomically corrected view) – showing both mitral and tricuspid valves opening into a single ventricle of LV morphology. A rudimentary RV type of ventricle is also visualized



**Figure 4:** (a) and (b) shows the pulmonary vegetation suggesting pulmonary endarteritis



**Figure 5:** CT scan of the brain showing bilateral cerebral abscesses

so with cyanotic congenital heart disease (CCHD).<sup>[3]</sup> Right-sided infective endocarditis is rare by virtue of the low hemodynamic pressure and lack of isolated or significant right-sided valvar deformities. In adults, right-sided infective endocarditis usually follows prolonged intravenous catheterization or intravenous drug-abuse, affecting the tricuspid valve alone or in combination with the pulmonary valve. A similar valvar involvement also occurs in infants and in children, but here congenital cardiac anomalies with their accompanying interventions are important pre-disposing factors.<sup>[4]</sup>

Pulmonary endarteritis as an entity of right-sided endocarditis is rare, even in patients of congenital heart disease.<sup>[2]</sup> Only few reports of pulmonary endarteritis are available. Most of the reported pulmonary endarteritis cases have been associated with a patent ductus arteriosus (PDA). Endarteritis is a rare event in PDA.<sup>[5]</sup> There are less than 30 reported cases of pulmonary endarteritis associated with PDA in the adult, and overall incidence has dramatically decreased over the last 30 years.<sup>[6,7]</sup> Bilge *et al.*, in their case report of pulmonary endarteritis in PDA, underlines the importance of echocardiography in not only making this rare diagnosis but also as an effective means of following up such a case.<sup>[8]</sup> Apart from PDA, a single report exists of pulmonary endarteritis occurring after anatomical correction of complete transposition of the great arteries.<sup>[9]</sup> To the best of our knowledge and available resources, our case report of pulmonary endarteritis in a child of single ventricle is probably the first such so far.

The onset of pulmonary endarteritis and the formation of vegetation can be explained by several factors,

including – an obstacle of a valve or bloodstream jet and venturi effect, the formation of a platelet fibrin thrombus, transient bacteremia, and adhesion factor for infection.<sup>[10]</sup> In the present case, an untreated infection in the child could have led to multiple episodes of bacteremia. The presence of multiple, bilateral cerebral abscesses is a corroborative evidence to the bacteremia. The pulmonary artery in this case arose from the main ventricle (morphological double-inlet LV) and was hypoplastic. A valvular pulmonary stenosis also existed. Turbulent blood flow distal to the stenotic lesion may increase the risk of developing infective endarteritis in an already hypoplastic pulmonary artery. The intima is also damaged in these areas, predisposing to formation of platelet fibrin thrombus, and thus making the region more susceptible to colonization by pathogens.<sup>[11]</sup>

Congenital heart disease (CHD) has a reported incidence of 9596/million live births (0.95%), out of which 14.5% (1391/million live births) have cyanotic congenital heart disease (CCHD). Single ventricle (106/million live births) accounts for 7.6% of CCHDs and 1.1% of all CHDs.<sup>[12]</sup> Single ventricle, therefore, is uncommon. Meticulous full-segmental echocardiographic approach is required to diagnose and delineate the specific type of single ventricle. Our case, a double inlet left ventricle with left-sided subaortic hypoplastic right ventricle (Van Praagh’s A-III type – Single Ventricle), is the most common variety of single ventricle.<sup>[13]</sup> Single ventricle is a complex CCHD and is considered a high risk condition for an infective endocarditis. Trans-thoracic echocardiography (TTE) is more sensitive in the pediatric population than in the adult population for detection of vegetation. TTE is more likely to identify vegetations in children with normal anatomy or isolated valvular pathology than in those with complex CCHD.<sup>[14]</sup> The importance of a carefully-performed TTE or trans-esophageal echocardiography is, therefore, essential in a complex CCHD like single ventricle states.

Cyanotic heart disease accounts for 12.8%-69.4% of all cases of brain abscesses, with the incidence being higher in children.<sup>[15]</sup> The incidence of brain abscess in patients with cyanotic heart disease has been reported to range between 5 and 18.7%.<sup>[16]</sup> In our case, bacteremia from an unknown source could have bypassed the pulmonary circulation by entering the double-inlet LV (the dominant ventricle) and moved through the aorta and seeded in the brain. Patients with cyanotic heart disease could have low-perfusion areas in the brain due to chronic severe hypoxemia and metabolic acidosis as

well as increased viscosity of blood due to secondary polycythemia. These low-perfusion areas commonly occur in the junction of gray and white matter, and they are prone to seeding by microorganisms that may be present in the bloodstream.<sup>[17]</sup> The advent of CT scans and their use in the management of these abscesses has resulted in a fourfold decrease in the mortality rate in patients with brain abscesses, secondary to cyanotic heart disease - from 40%-60% in the pre-CT era to ~10%.<sup>[18]</sup> The threshold for CT scans in a CCHD should be low, and regular CT scans should be obtained to monitor the size of the abscess.

In conclusion, this case report aims to highlight the considerable and varied morbidity associated with cyanotic congenital heart diseases (CCHD). The presence of a relatively common infectious complication (cerebral abscesses) and a rare infectious complication (pulmonary endarteritis) in the same child with a CCHD (single ventricle) validates the need for a heightened clinical awareness and a prompt and meticulous investigational approach for diagnosis and therapy in this group of patients.

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