

# Pediatric Stroke: Neurological Sequelae in Uncorrected Tetralogy of Fallot

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## Abstract

Pediatric stroke is an uncommon entity. A risk factor is present in almost half of the children at the time of stroke. The most common cause of stroke in children is probably congenital heart disease. Other risk factors include sickle cell disease, infections, and various prothrombotic conditions. We present a case of a 3-year-old male child, who was previously diagnosed with Tetralogy of Fallot and presented with left-sided weakness and one episode of generalized tonic-clonic seizures.

**Keywords:** Africa, Congenital heart disease, Pediatric stroke, Stroke, Tetralogy of Fallot

## Introduction

The estimated incidence of ischemic stroke in children older than 28 days of life is variable,<sup>[1-3]</sup> but according to a prospective population study, it averages 13/100, 000 for all strokes, 7.9/100, 000 for ischemic strokes and 5.1 for hemorrhagic strokes.<sup>[2]</sup>

Systemic arterial circulation can be the source of emboli; however, mechanism via paradoxical embolus has been well-established.

Long-standing cyanotic lesions cause polycythemia and anemia, which increase the risk of thromboembolism and cerebral infarction.<sup>[4]</sup> Polycythemia not only increases the risk but can also mimic computed tomography (CT) sign of cerebral venous thrombosis. Magnetic resonance imaging (MRI) is necessary to confirm the diagnosis.<sup>[5]</sup>

## Case Report

A 3-year-old male child was brought to the hospital with complaints of weakness of the left upper and lower limbs

and one episode of generalized tonic-clonic seizures associated with up rolling of eyeballs and frothing from the mouth. Child was a diagnosed case of Tetralogy of Fallot following history of recurrent chest infections in infancy. He had two episodes of vomiting and five to six episodes of loose motions per day since 2 days. There was no history of surgical intervention (Informant – Mother). Lab investigations revealed PCV-46%, Hb-14.1 gm/dL, mean cell volume-61.33 cumm (80-90)-microcytic, TLC-13,400 cumm, and neutrophils – 74%.

CT scan was performed on emergency basis [Figures 1-3]. For further evaluation, MRI with magnetic resonance (MR) venography was also performed the next day [Figures 4-7].

## Discussion

Stroke and brain abscess are the most common neurological complications of uncorrected congenital heart disease.<sup>[6]</sup> Corrected as well as uncorrected cases of Tetralogy of Fallot are associated with increased risk of developing pulmonary thrombosis, venous thrombosis, cerebral embolism, and infective endocarditis.<sup>[6]</sup>

Various causes of stroke have been recognized in children with cyanotic heart disease including thromboembolism, prolonged hypotension, and polycythemia.<sup>[6]</sup> Iron-deficiency anemia and dehydration are other factors that hasten the formation of thrombus in these patients.<sup>[7]</sup> Cerebral emboli originate either in the systemic arterial circulation or in the venous circulation, that is, lower extremity veins or pelvic veins.

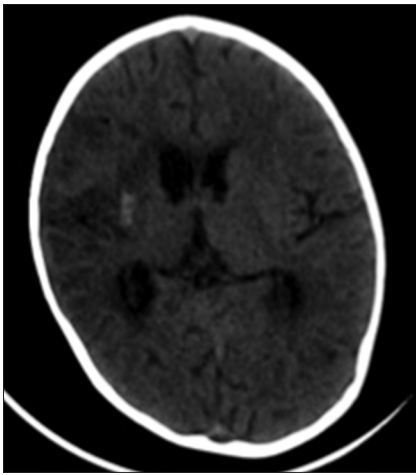
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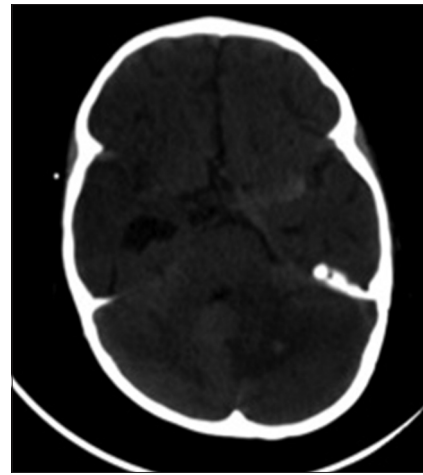


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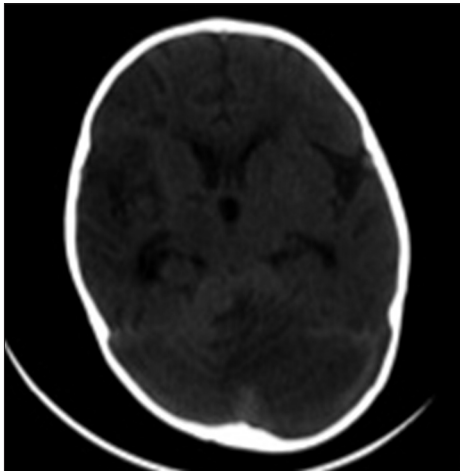
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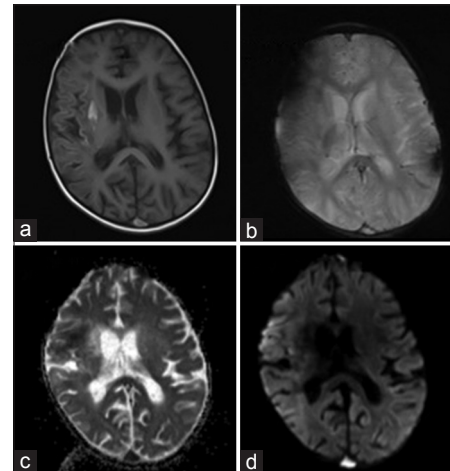
**Figure 1:** Plain CT scan image showing hyperdense area in the right lentiform nucleus with surrounding ill-defined hypodense area



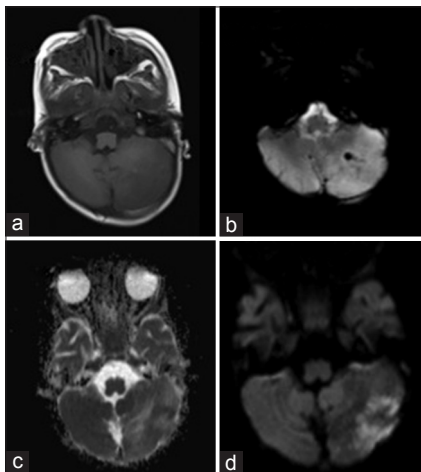
**Figure 2:** Plain CT scan image showing diffuse ill-defined hypodense area in the left cerebellar region, with hyperdense focus within it, causing compression and displacement of the fourth ventricle. Dilatation of the contralateral temporal horn of the lateral ventricle is noted



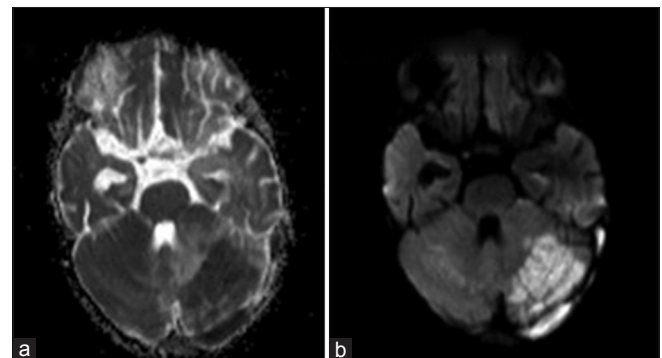
**Figure 3:** Hyperdensity seen in the region of superior sagittal sinus and left transverse sinus



**Figure 4:** Hemorrhagic infarct in right lentiform nucleus (subacute). (a) Axial T1WI shows a hyperintense area in the right lentiform nucleus. Also note subtle hyperintensity in the superior sagittal sinus. (b) Lesion does not bloom on gradient sequence. (c) On ADC lesion appears bright. (d) No restriction seen on diffusion sequence



**Figure 5:** Hemorrhagic infarct in left cerebellar hemisphere. (a) Axial T1WI shows ill-defined hypointense area with few hyperintense foci in the left cerebellar hemisphere. Also hyperintensity is noted in the region of left transverse and sigmoid venous sinus. (b) On gradient sequence, few central areas of blooming are noted. (c) On ADC lesion appears hypointense. (d) Restriction is noted at the corresponding level on diffusion sequence



**Figure 6:** Hemorrhagic infarct in left cerebellar hemisphere. (a) ADC image cranial to previous sections shows diffuse hypointense area in the left cerebellar hemisphere. (b) Restriction is noted at the corresponding level on diffusion sequence

Stroke due to cerebral arterial thrombosis has been attributed to the secondary erythrocytosis of cyanotic heart disease, which is pathologically different from primary erythrocytosis of polycythemia vera. An association between elevated hematocrit and thrombosis has been established in polycythemia vera. However, no association has been established between elevated hematocrit and secondary erythrocytosis in case of cyanotic heart disease.<sup>[7]</sup>

Another cause of cerebral infarction in children less than 4 years of age is thrombosis of intracranial dural sinuses or cerebral veins usually in association with iron-deficiency anemia. Superior sagittal sinus, transverse sinus, great vein of Galen, and meningeal veins are the common sites of thrombosis in decreasing order of precedence. Intracranial hypertension may be the only manifestation if the thrombosis is limited to superior sagittal sinus or transverse sinus. Partial or complete recovery is possible even with a severe initial presentation emphasizing the need for early diagnosis and treatment; CT scan is often the first investigation performed on an emergency basis.<sup>[8]</sup>

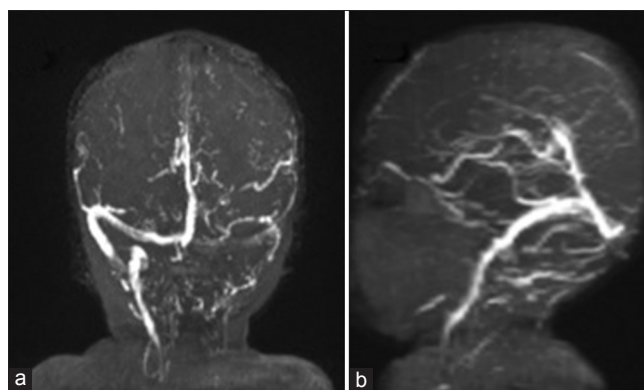
Unenhanced CT scan demonstrates venous thrombosis as linear hyperdensities in the expected locations of the dural venous sinuses and cortical veins. As the thrombus becomes less dense, contrast-enhanced CT scan demonstrates the “empty delta” sign, that is, a filling defect in the posterior part of the sagittal sinus.

Increased attenuation of cerebral veins and sinuses is also noted in cases of polycythemia. New and Aronow<sup>[9]</sup> discovered a linear relationship between CT attenuation values and the hematocrit of whole blood preparations. The CT attenuation of hemoglobin was found to be largely due to its protein content. The iron in blood contributed only about 7% of the total attenuation. They also demonstrated that the attenuation of retracted clot was approximately 90%, twice the attenuation of normal intravascular blood (hematocrit level of 43).

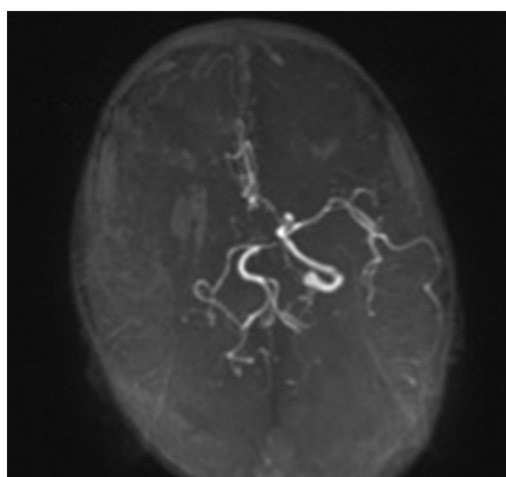
Thus, polycythemia is not only a risk factor for development of venous thrombosis, but also a mimicker of the same and if these two conditions coexist, as in the case of Tetralogy of Fallot, the differentiation between the two is only possible by contrast-enhanced CT scan or on MRI.<sup>[5]</sup>

In our case, child has extensive venous thrombosis involving the superior sagittal sinus, left-sided venous sinuses, and the left internal jugular vein leading to multiple hemorrhagic infarcts [Figure 7]. Nonvisualization of right internal carotid artery from its cervical portion can be attributed to embolic phenomenon [Figures 8 and 9]. PCV was 46% – mildly elevated level. Hyperdensity in the region of dural venous sinuses was confirmed to be thrombosis on MR venography.

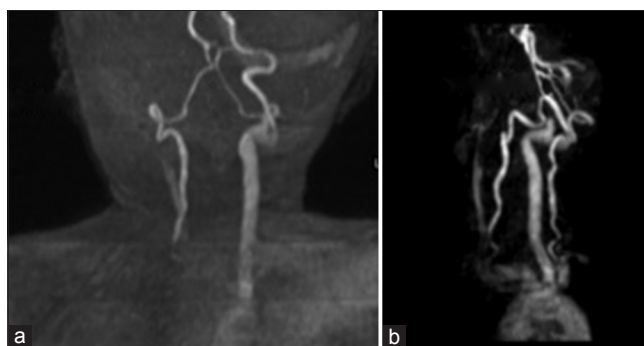
Various options for treatment in children with venous sinus thrombosis include standard or low molecular weight heparin



**Figure 7:** Venous sinus thrombosis. (a) Coronal, (b) Sagittal two-dimensional TOF images demonstrating thrombosis of superior sagittal sinus, left transverse sinus, left sigmoid sinus, and left internal jugular vein



**Figure 8:** Three-dimensional TOF image depicting nonvisualization of right internal carotid artery and its branches



**Figure 9:** (a) Coronal MIP3D TOF image of neck vessels and (b) three-dimensional TOF image of aortic arch and its branches show nonvisualization of the cervical portion of the right internal carotid artery

for 7-10 days, followed by oral anticoagulants for 3-6 months. Though anticoagulation is controversial in children, treatment with heparin is probably safe and beneficial for these children, even for those with intracranial hemorrhage.<sup>[8]</sup>

The clinical practice to phlebotomize patients with polycythemia in order to maintain hematocrit more than

65% has been long followed. Alternative treatment is volume replacement and low-dose iron therapy as repeated phlebotomy can cause iron deficiency leading to microcytic erythrocytes which increases the whole blood viscosity ultimately increasing rather than decreasing the risk for stroke.<sup>[7]</sup>

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