Acute ischemic stroke in a child with cyanotic congenital heart disease due to non-compliance of anticoagulation

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BACKGROUND: Stroke is a common presentation in geriatric patients in emergency department but rarely seen in pediatric patients. In case of acute ischemic stroke in pediatric age group, management is different from that of adult ischemic stroke where thrombolysis is a good option.

METHODS: We report a case of a 17-year-old male child presenting in emergency with an episode of acute ischemic stroke causing left hemiparesis with left facial weakness and asymmetry. The patient suffered from cyanotic congenital heart disease for which he had undergone Fontan operation previously. He had a history of missing his daily dose of warfarin for last 3 days prior to the stroke.

RESULTS: The patient recovered from acute ischemic stroke without being thrombolized.

CONCLUSION: In pediatric patients, acute ischemic stroke usually is evolving and may not require thrombolysis.

KEY WORDS: Acute ischemic stroke; Congenital heart disease; Single ventricle; Fontan operation; Warfarin; Thrombolytic

INTRODUCTION

Stroke has long been associated with aging and co-morbidities owing to atherosclerosis. As of now stroke is not a common disease in the pediatric population. Because of lack of awareness about the incidence¹ of stroke in the pediatric age group,² such patients usually present late in the emergency department. By the time, the patient is brought to the hospital, either the window period of thrombolysis (in case of acute ischemic stroke) has usually passed or the patient has recovered from the focal neurological deficit, if any.

As per the definition of stroke given by the WHO, "Stroke is rapidly developed clinical signs of focal disturbance of cerebral function, lasting for more than 24 hours or leading to death, with no apparent cause other than vascular origin". This is not the usual form of stroke seen in children. In case of children, stroke can even last for less than 24 hours² and can still show changes on neuroimaging.³

In children, acute ischemic stroke is commonly associated with underlying conditions like congenital or acquired heart disease and sickle cell disease. Cyanotic congenital heart diseases with right-to-left shunt are particularly prone to cause acute ischemic stroke. Though

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stroke is more common in children with uncorrected congenital heart disease, it occurs even after the correction of the defect. The following is a case report of a child who presented to the emergency department with acute ischemic stroke with underlying cyanotic congenital heart disease namely, single ventricle.

Case report
A 17-year-old male child presented to the emergency department with complaints of left sided weakness and facial asymmetry. The patient woke up early morning because of headache which was followed by a bout of vomiting. Simultaneously, the patient noticed that he was unable to move his left upper and lower limbs along with facial asymmetry on the same side. He presented to the emergency department of our hospital within a couple of hours with complaint of inability to move the left upper limb with residual weakness in the left lower limb and left facial weakness as well as asymmetry. He did not have any complaint of fever, loss of consciousness, urinary incontinence or seizure.

On primary survey, the patient was hemodynamically stable with a borderline hypertension (BP=140/100 mmHg) and a GCS score of 15 (E4 M6 V5) with dilated left pupil but both of the pupils were equally reacting to light. He was unable to move his left upper limb and had facial weakness on the left side. On secondary survey, the patient had normal higher mental function. On motor system evaluation the patient had power of 0/5 in the left upper limb, 5/5 in the left lower limb (recovered from 0/5), 5/5 in the right upper limb and 5/5 in the right lower limb. On reflex evaluation, his left plantar was extensor while the right plantar was flexor. The rest of the systemic evaluation was normal. He had a NIH stroke score of 2.

The patient had a history of cyanotic congenital heart disease namely single ventricle, for which Fontan operation was done 10 years ago. The patient had been taking warfarin and enalapril daily, but he missed his daily dose of warfarin for last 3 days. He was neither non-diabetic nor non-hypertensive with no underlying sickle cell disease.

On imaging, NCCT of the brain was normal while MRI of the brain was suggestive of acute non-hemorrhagic infarcts involving the right sylvian cortex and adjoining the right frontal gray and white matter. MRI of the brain was suggestive of a normal study. On blood investigation, INR of the patient was found to be 1.45. Echocardiography showed post-operative fenestrated Fontan with a small right-to-left shunt across the fenestration. Normal left ventricular function was noted with moderate mitral regurgitation. Normal functioning was noted of the inferior vena cava to the pulmonary artery and the superior vena cava to the pulmonary artery conduits.

The patient had a complete recovery of neurological deficits within 4 hours and thrombolysis was deferred in view of recovering stroke. He was given low molecular weight heparin in the emergency department. Later he was admitted to the ward for inpatient care.

DISCUSSION
In India, where emergency medicine is still a new branch, presentation of pediatric stroke patients to the
department and their early recognition is very rare. In this case, the parents of the patient were regularly followed up by the pediatric cardio-thoracic surgeon who had operated on him a decade ago. When the surgeon was contacted regarding the onset of symptoms, the patient was immediately referred to the pediatric neurologist, who advised the parents to immediately take the patient to the emergency department. In this patient, the early presentation of acute ischemic stroke and the early recovery were seen. The association of the recovery of neurological focal deficits with ischemic changes on MRI in this patient was different from that of the patient with transient ischemic stroke, which usually has a recovery of neurological focal deficits but no ischemic changes are seen on neuroimaging.

In India, the incidence of cyanotic congenital heart disease is far more common than sickle cell disease, especially in the northern part. Thus, the underlying cause for acute ischemic stroke is commonly cyanotic congenital heart disease. According to the report\(^1\) of the American Stroke Association, complex congenital heart lesions with right-to-left shunting and cyanosis are particularly prone to cause stroke. Although stroke is more common among children with uncorrected congenital heart lesion, surgical correction of the lesion reduces the risk of stroke but does not completely eliminate it. Thus the prolongation of anticoagulation therapy could be needed in such population. Warfarin is commonly used for this purpose in children with congenital heart disease, even after correction.\(^2\) The use of thrombolytic agents in a setting of acute ischemic stroke in children has not been well recognized so far. The delayed presentation and recognition preclude the use of thrombolytics in children with acute ischemic stroke, and thus insufficient data are available on the effectiveness of thrombolytic therapy in children. In this report, thrombolytic therapy was deferred even though the presentation of the patient was within the window period. The reason for this was the continuing recovery of the patient after the onset of symptoms. Hence the risk of using thrombolytic therapy like intracranial hemorrhage outweighed the benefit of its use. And the patient received low molecular weight heparin once maximum recovery was noticed within the window period.

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**REFERENCES**


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