

Acute Stroke Syndrome in Pediatric Age Group: A ReviewAKM Kamruzzaman¹, Most. Manzuara Khatun², MH Haidary³¹Public Health and Child Specialist, ²Assistant Professor, Department of Anatomy, Islami Bank Medical College, Rajshahi, ³Professor and Head, Department of Pediatrics, Islami Bank Medical College, Rajshahi.**Abstract**

Neurological deficits in the pediatric age group are much rarer than in adults. Neurological deficits caused by cerebrovascular diseases are defined as stroke, whereas conditions manifesting with neurological deficits without underlying cerebrovascular diseases are referred to as stroke mimics. Stroke is relatively rare in children, but can lead to significant morbidity and mortality. Understanding that children with strokes present differently than adults and often present with unique risk factors will optimize outcomes in children. Despite an increased incidence of pediatric stroke, there is often a delay in diagnosis, and cases may still remain under or misdiagnosed. Clinical presentation will vary based on the child's age, and children will have risk factors for stroke that are less common than in adults. Currently, there is no consensus on the classification, evaluation, outcome measurement, or treatment of perinatal and childhood stroke. Pediatric stroke registries are needed to generate data regarding risk factors, recurrence and outcome. The impact of maternal and perinatal factors on risk and outcome of neonatal stroke needs to be studied. This information is essential to identifying significant areas for future treatment and prevention.

Key words: Acute stroke syndrome, Cerebrovascular disease

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Introduction

Acute stroke syndrome is a neurological injury caused by the occlusion or rupture of cerebral blood vessels. Stroke can be ischemic, hemorrhagic or both. Ischemic stroke is usually caused by arterial occlusion, but it may also be caused by venous occlusion of cerebral veins or sinuses. Stroke is relatively rare in children, but can lead to significant morbidity and mortality. Roughly 10-25% of children with a stroke will die, up to 25% of children will have a recurrence and up to 66% will have persistent neurological deficit or develop subsequent seizure disorders, learning or developmental problems.^{1, 2, 3}

Acute ischemic stroke accounts for half of all strokes in children, in contrast to adults in whom 80-85% of all strokes are ischemic.^{4, 5}

There is widespread belief that strokes don't happen to children. This may be due to inadequate investigation facility and very little data base.

Early recognition of acute stroke syndrome may lead to more rapid neurological consultation, imaging, treatment and improved outcomes. Many studies on acute stroke syndrome and pediatric stroke are conducted in different parts of the globe. But studies in acute stroke syndrome in pediatric age group in our country are very lacking. In this article we do review the epidemiology, risk factors, causes and clinical presentation of acute stroke syndrome in pediatric age group so that inadequate diagnosis of acute stroke syndrome related morbidity and mortality could be minimized.

Epidemiology

A stroke or cerebrovascular accident (CVA) in children is typically considered to be a rare event. The reported incidence of combined ischemic and hemorrhagic pediatric stroke ranges from 1.2 to 13 cases per 100,000 children under 18 years of age per year. However, incidence of stroke in children is likely more than we think and it is thought to be frequently undiagnosed or misdiagnosed. Pediatric stroke is more common in

Address for Correspondence : AKM Kamruzzaman, Surveillance Medical Officer- WHO, Civil Surgeon Office, Rajshahi, Bangladesh.

boys than girls and appears to be predominance in black children.^{1, 7-14}

Risk factors and causes of childhood strokes

Strokes in adults often can be blamed due to high blood pressure, high cholesterol level, a history of smoking, too much alcohol and obesity. The majority of signs and symptoms of acute stroke syndrome are nonspecific and can be easily attributed to other causes. Multiple risk factors are often present in as many as 25% of children with stroke, which means further investigations are warranted even when one risk factor has been identified.^{13, 14}

In contrast, children's strokes are often caused by:

- Birth defects (cardiac, vascular)
- Birth trauma
- Perinatal asphyxia
- Syndromic and metabolic disorder
- Infections (e.g., meningitis, encephalitis)
- Blood disorders such as sickle cell disease
- Trauma

Classification and presentation of pediatric strokes

1. Arterial ischaemic stroke

The incidence of arterial ischaemic stroke in childhood is for about half of all strokes in children. The most common presentation of arterial ischaemic stroke (AIS) in childhood is hemiparesis/ hemiplegia attributable to the relatively frequent involvement of the middle cerebral artery. Other less common presenting features include altered mental status and focal neurological signs, such as aphasia and visual disturbance. Stroke in the posterior lobe is rare and when it occurs it is usually associated with vertebral artery dissection, presenting with ataxia, vertigo or vomiting.

2. Moyamoya disease/syndrome

Moyamoya disease is a progressive occlusive vasculopathy. On angiography, the characteristic appearance of dilated lenticulostriate perforating vessels is described as a puff of cigarette smoke or moyamoya in Japanese. Moyamoya can either be primary or idiopathic (in which case it is referred

to as moyamoya disease) or secondary related to underlying disorders such as Down's syndrome, sickle cell disease, neurofibromatosis type 1 or cranial radiotherapy (in which it is referred to as moyamoya syndrome). The clinical symptoms include headache, transient ischaemic attack, AIS, intracranial haemorrhage or silent cerebral infarction.

3. Metabolic stroke

Inborn metabolic disorders are generally caused by spontaneous mutations or hereditary gene defects resulting in biochemical alterations involving one or more metabolic pathways. The clinical features are non-specific, variable and may at times mimic stroke.

4. Mitochondrial encephalopathy with lactic acidosis and stroke like episodes

Patients with mitochondrial encephalopathy with lactic acidosis and stroke-like episodes (MELAS) syndrome are usually healthy at birth. Later, the affected children exhibit delayed growth, episodic vomiting, seizures and recurrent cerebral injuries resembling stroke. These stroke-like events, probably as the result of proliferation of dysfunctional mitochondria in the smooth muscle cells of small arteries may give rise to either permanent or reversible deficits.

5. Hemorrhagic stroke

Hemorrhagic stroke is rare in children. The reported incidence is approximately 1.2 per 100000 children per year. Hemorrhagic stroke more commonly present with headache or altered level of consciousness and vomiting. Traumatic head injury, non-accidental injury, bleeding disorders (hemophilia), arteriovenous malformations and hemorrhagic neoplasms are some of the leading causes of hemorrhagic stroke in children.

6. Venous stroke/thrombosis

Cerebral sinovenous thrombosis (CSVT) occurs in 0.41 per 100000 live-born infants and 40% of pediatric patients with venous thrombosis have venous infarction. Seventy percent of venous infarctions are haemorrhagic. Clinically, infants most commonly present with seizures and diffuse neurological symptoms. Focal neurological signs,

headache and alteration of consciousness are typical symptoms in older children.

At-A-Glance Signs & Symptoms of Childhood Stroke Syndromes*

Acute Arterial Ischemic Stroke	
Medical description	Lay description
Hemiparesis	Weak arm or leg, facial droop, paralyzed one side
Aphasia	Stopped speaking, talking nonsense, won't follow command
Dysarthria	Slurred speech
Hemisensory loss	Numbness, tingling on one side of the body
Acute Hemorrhagic Stroke	
Medical description	Lay description
Severe Headache	Worst headache of my life
Sudden loss of consciousness	Collapsed, hard to wake up
Hemiplegia	Paralyzed on one side, eyes going to one side, face drooping

Use the following tool to help you think fast to diagnose childhood stroke syndrome and take decision immediately:

F. Face	Ask the child to smile. Does one side of the face droop?
A. Arms	Ask the child to raise both arms. Does one arm drift downward?
S. Speech	Ask the child to repeat a simple sentence. Are the word slurred?
T. Time	If the child shows any of those symptoms, time is important. The hospital fast as brain cell are dying.

Case History:

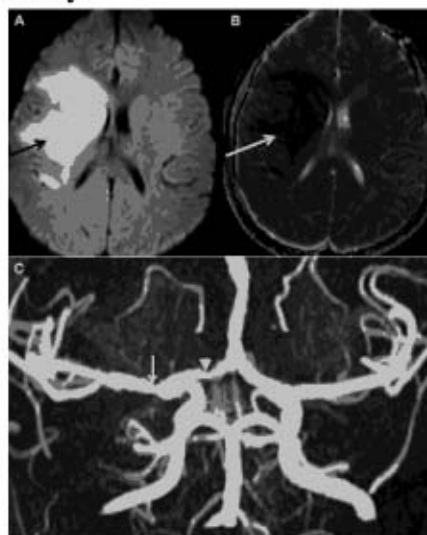


Figure 1 : MRI of 1st case

A 14 year old patient presented with sudden onset of left-sided weakness and facial palsy. MRI brain shows : A diffusion-weighted imaging (DWI) and B apparent diffusion coefficient (ADC) map. Hyperintensity on DWI (black arrow) and corresponding hypointense signal on ADC map (white arrow) are compatible with restricted diffusion over the right parietal region. Features are suggestive of an acute infarct. C MRI angiogram of the circle of Willis using the time of

flight technique reveals short segments of severe stenosis in the proximal M1 segment of the right middle cerebral artery (white arrow) and distal A1 segment of the right anterior cerebral artery (white arrowhead).

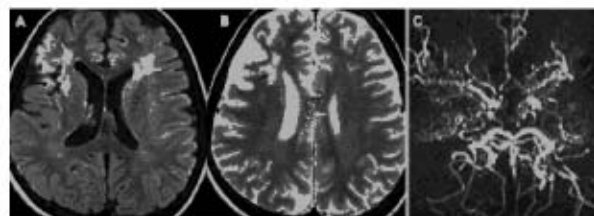


Figure 2 : MRI of 2nd case

A 13 year old boy presented with recurrent headache and repeated episodes of bilateral lower limb weakness. MRI shows : A Fluid-attenuated inversion recovery image shows hyperintensities in the bilateral frontal periventricular white matter (white arrows). B T2-weighted axial image shows prominent serpiginous collaterals in the parasagittal region (white arrow). C Magnetic resonance arteriography confirms absent bilateral internal carotid arteries associated and severe middle cerebral artery (MCA) stenosis (arrowhead). Overall features are consistent with moyamoya disease.

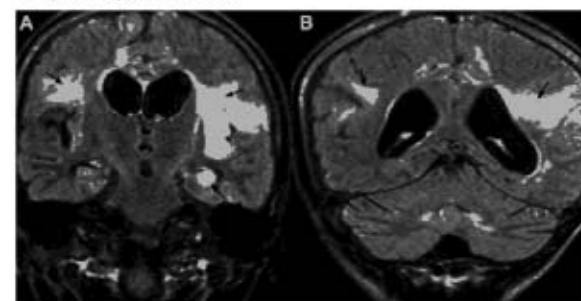


Figure 3 : MRI of 3rd case

A 4 year old girl presented with left-sided weakness. The cause of infarcts was not apparent at presentation but she was subsequently diagnosed to have mitochondrial myopathy, encephalopathy, lactic acidosis, and stroke syndrome by biochemical tests. No basal ganglia lesions were detected in this case. MRI shows : A and B Fluid-attenuated inversion recovery coronal images show multiple T2 hyper intensities in the bilateral cerebral hemispheres involving the bilateral insular cortices, left hippocampus and

bilateral corona radiata (black arrows).

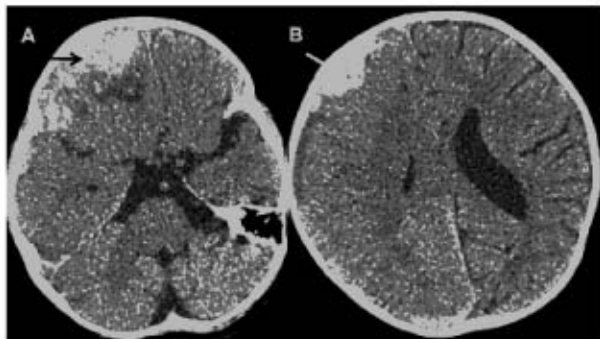


Figure 4 : CT scan of brain of 4th case

A 15 year old boy presented with sudden onset of disorientation and right-sided weakness. CT scan shows : unenhanced images A showing right frontal lobe haemorrhage and perilesional white marrow oedema (black arrow) and B showing right epidural haematoma (white arrow). The boy later was found to have haemophilia A.

Conclusion

Acute stroke syndrome in pediatric patient is a rare but potentially devastating disease. Children have a better ability to heal because of the greater plasticity or flexibility of the child's nervous system and brain. A child's brain is still developing; therefore it may have a greater ability to repair itself. With the help of physical and speech therapy, most childhood stroke survivors recover the use of their arms, legs and speech. However, because the incidence is still low in comparison to adult strokes, and children are distinctly different from adults, it remains a great challenge to create evidence based diagnostic and treatment guidelines. As there is widespread decision disparity and low incidence of this disease, future review or research needs nationally and internationally. Until then, acute stroke should remain a strong consideration in children with concerning signs and symptoms and significant risk factors, and best available evidence should be utilized in providing optimal medical care.

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