

# CEREBRAL BYPASS

## An Innovative Treatment for Arteritis



INSTITUTE OF NEUROSURGERY  
&  
DEPARTMENT OF PICU



# CASE 1



- ❑ 1 year old girl -recurrent seizure, right side limb weakness, excessive cry and irritability.
- ❑ Background: normal birth and development till 3 months
- ❑ Developed GTCS at 4 months of age
- ❑ She subsequently developed weakness from 6 months of age
- ❑ Right focal seizures - followed by which she developed right hemiplegia which marginally improved .

## *Contd...*



- ON EXAMINATION: Hemodynamically Stable  
Other systems within normal limits
- NEUROLOGICAL EXAMINATION :  
Child – conscious with GCS 15/15  
Right hemiparesis(+), Irritable  
Child was evaluated for thrombotic and vascular etiology

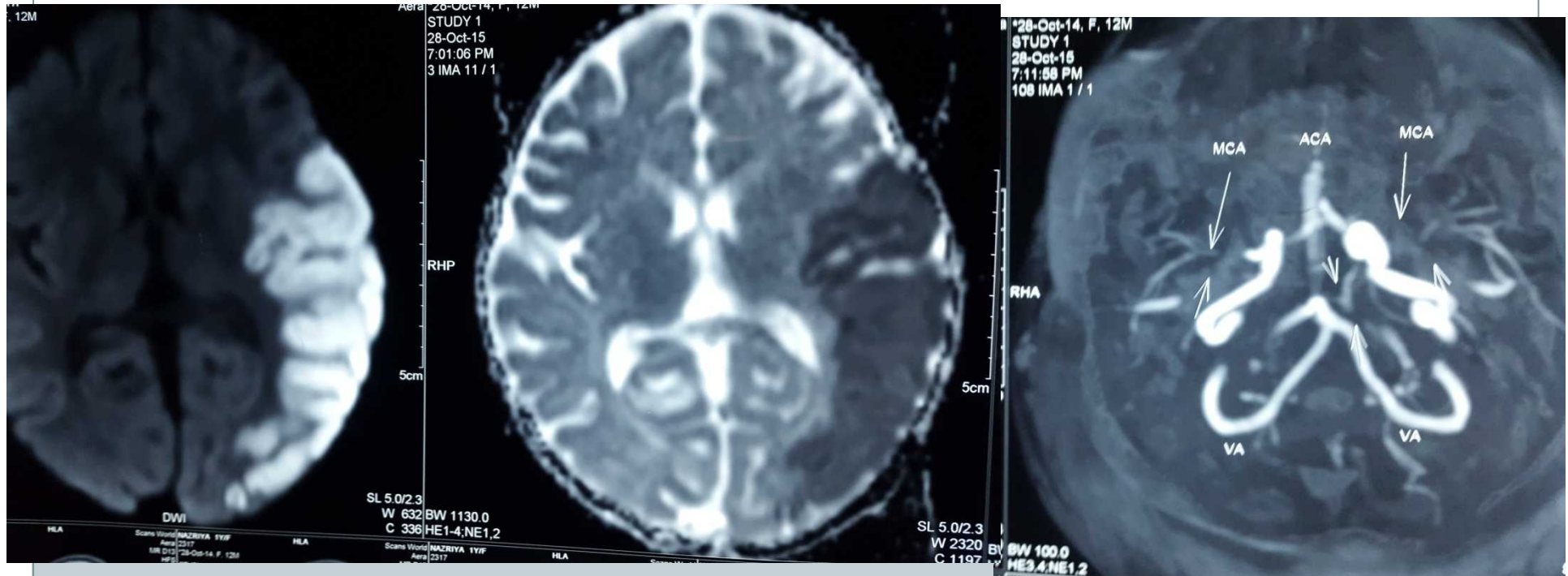
# Lab Evaluation



## Work Up for Thrombovascular etiology

- ❑ First line investigations-  
CBC, Coagulation profile, smear - Negative
- ❑ Second line investigations
  - ❑ Protein C and S - Negative
  - ❑ Antithrombin III - Negative
  - ❑ APC - Negative
  - ❑ Plasma Homocysteine levels - Negative

# Neuroimaging



## INVESTIGATIONS:

CT Angio : Bilateral MCA stenosis with bilateral basal ganglia neovasculature  
S/o Moya moya disease along with temperoparietal infarct



- Puff of smoke – though well visualized in conventional angiography can still be visualized in CT angiography

## CASE 2



- ❑ 3 year old female child brought with complaints right focal seizures associated with episodes of right side weakness.
- ❑ She was on Carbamazepine for seizure control.
- ❑ Background: normal birth and development
  - ❑ She had recurrent seizures twice in October and Thrice in November 2015

She was admitted for further management.

## *Contd...*



- ON EXAMINATION: Haemodynamically stable
- Neurological Examination

Right facial nerve palsy

Cognitive function-Normal

Child conscious

Right hemiparesis(+)

Child was evaluated for thrombotic and vascular etiology - negative



# Neuroimaging



CT Angiogram- bilateral ICA stenosis with puff of smoke appearance left > right with multiple old infarcts

# *Diagnosis*



*MOYA MOYA disease*

# Surgery Done



Cerebral revascularization technique / Cerebral bypass

LEFT STA – MCA ANASTOMOSIS

&

ENCEPHALO-DURO-MYO-SYNANGIOSIS  
(EDAMS)

# *Moya Moya Disease*



- Moyamoya disease - cerebrovascular arteriopathy of unknown origin
- Progressive stenosis and, ultimately, occlusion of the distal intracranial internal carotid arteries (ICA) and the proximal branches of the anterior and middle cerebral arteries.
- Collateral vessels develop from the leptomeninges as well as both the external and intracranial ICAs for reperfusion.
- Collaterals lead to the characteristic angiographic appearance likened to a puff of smoke.

# *Moya Moya Disease*



- In 1957 by Takeuchi and Shimizu as a case of “hypoplasia of the bilateral internal carotid arteries.”
- In 1969 when Suzuki and Takaku coined the term “moyamoya” signifying “something hazy, like a puff of cigarette smoke” to describe the angiographic appearance.

# Moyamoya disease versus syndrome



- Patients with the characteristic vasculopathy who also have well **recognized associated conditions** are categorized as **Moyamoya syndrome**.
- Patients with **no known associated risk factors** are said to have **Moyamoya disease**.

## Associated characteristics and conditions

Common	50-75
Angiographic findings of moyamoya without other disease	
Asian heritage	
Less common (moyamoya syndrome)	10-20
Sickle cell disease	
Neurofibromatosis type 1	
Cranial therapeutic irradiation	
Down's syndrome	
Rare (moyamoya syndrome)	<10
Congenital cardiac anomaly	
Renal-artery stenosis	
Giant cervicofacial hemangiomas	
Hyperthyroidism	

# *Clinical presentation*



- Adults present - intracranial hemorrhage (~46%).
- Children present typically with signs and symptoms of **cerebral ischemia secondary to TIAs and/or cerebral infarctions**.
- Ischemia to the frontal, parietal, and temporal lobes are associated with hemiplegia or paresis, sensory loss or parasthesias, aphasia, and/or cognitive impairments

*(Both our cases presented with infarcts with focal deficits and had cognitive impairments)*

# Natural History



- Slow progression with intermittent events, to rapid neurological and cognitive decline, with overall mortality rates being high.
- Long-term outcome of Moyamoya disease is poor.
- 66% patients have symptomatic progression over a 5-year period following diagnosis



# *Revascular surgery*



Revascularization surgery helps in maintaining the blood supply to distal portions and prevent recurrent ischemic attacks

## Revascular surgery

1. Direct surgery
2. Indirect surgery
3. Combined surgery

# *Direct surgery*



## *Direct superficial temporal artery-to-middle cerebral artery bypass (STA-MCA)*

- Superficial temporal – Middle cerebral artery
- Occipital artery – Middle cerebral artery.
- The major advantage - an immediate increase in blood flow to the ischemic brain
- Smaller arteries - at risk for ischemic events
  - Thrombosis

# *Indirect surgery*



Commonly employed Indirect surgery include:

- a. Encephalomyosynangiosis(EMS)
- b. Encephaloduroarteriosynangiosis(EDAS)
- c. Pial synangiosis
- d. Encephaloduroarteriomyosynangiosis( EDAMS)
- e. Omental flaps
- f. Cranial burr holes

In both the children Combined approach  
SCA-MCA + EDAMS

**Table 2** Children treated with moyamoya at three different hospitals

Variables	The Children's Hospital, Boston, Massachusetts	Stanford University, California	Johns Hopkins Hospital, Maryland
No. of patients (age range at time of surgery)	143 (0.5–21 years)	96 (1–17.9 years)	14 (0.1–13.9 years)
No. of surgical procedures	271	168	23
1. Direct	0%	76.2%	0%
2. Indirect	100%	23.8%	87%
3. Combined	0%	0%	13%
Moyamoya type			
1. Disease	66	80	8
2. Syndrome	77	16	6
Presenting symptoms			
1. Ischemic	67.8% stroke, 43.4% TIA	51%	85.7%
2. Hemorrhagic	2.8%	2.1%	0%
3. Headache	6.3%	44%	0%
4. Seizure	6.3%	NR	8.3%
5. Choreiform movements	4.2%	NR	0%
6. Incidental	4.2%	NR	8.3%
Immediate postoperative complications (<30 days)			
CVA	11	2	0
1. TIA (severe)	3	0	2
2. Hemorrhage	0	1	0

Childs Nerv Syst (2010) 26:1297–1308  
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SPECIAL ANNUAL ISSUE

## Moyamoya disease in children

David M. Ibrahim · Rafael J. Tamargo ·  
Edward S. Ahn

# Concerns and issues in the Post operative period



- The risk of stroke – during first 30 days after surgery.
- 96% - stroke free over the subsequent 5-year period.
- Preventing ischemic events - reduce pain, crying, and hyperventilation induced cerebral vasoconstriction, painless wound dressings and absorbable sutures.
- Avoid Hypotension, hypovolemia, hyperthermia, hypercapnia, and hypocapnia.
- Peri-operative period – Heparin + aspirin and long-term Aspirin thereafter.

# *Post Op Course in Hospital*



## Sedation and Analgesia to avoid agitation and crying

- Following procedure ,both children were extubated in OT and shifted to PICU for post op care.
- Sedation with Dexmedetomidine infusion & Trichlorofos.
- Analgesia with Morphine infusion
- Aspirin was given orally and continued long term.



- In Case I :
  - ICP monitoring was done using EVD –external ventricular drainage with intermittent surges being treated by opening the drain, letting out CSF and increasing the level of sedation
  - EVD was removed after 48 hours once sedation and analgesia were tapered and stopped.
- In Case II:
  - required more sedation to keep the child quiet
  - early feeding and oral sedation and analgesia

Both the children are doing fine – irritability decreased.

Sensory evoked potential checked in one child – normal.

# Take Home Messages



- MoyaMoya – rare disease and has morbid outcome
- Early referral and bypass surgery will help in preventing ischemic events and morbidity





- Dr Roopesh Kumar & Dr Suresh Bapu  
Senior Consultant – Institute of Neurosurgery  
SIMS Hospitals, Vadapalani - Chennai