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## **A rare entity with a rarer presentation: A case report on Dyke-Davidoff Masson syndrome presenting with status epilepticus**

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**D**yke-Davidoff-Masson syndrome (DDMS) was described initially by C G Dyke, L M Davidoff and C B Masson in 1933. It results from an insult to the growing brain in utero or early infancy, leading to loss of neurons, compromising the growth of the brain. Diagnosis is established with clinical and neuroimaging data. Patients may require physiotherapy, speech and occupational therapy in addition to the management of the seizures. Awareness among the pediatrician, the radiologist and the team leads to early recognition, appropriate timely management and hence, better long-term prognosis. A nine years old female born prematurely, presented thrice in her lifetime as a case of status epilepticus due to improper and inadequate use of prescribed medications. She had delayed mental development for her age. Upon contrast, CT evaluation, hemiatrophy of the right cerebral hemisphere with compensatory ex-vacuo ventriculomegaly and midline shift towards the right, ipsilateral thickening of the calvarium, decrease in size of the ipsilateral cranial fossa, unilateral overdevelopment of the frontal sinus and overaerated right petrous were noted. Capillary malformation was seen as spoke-wheel appearance of the capillaries in the right basal

ganglia, a novel finding in children with DDMS. She was diagnosed as a case of DDMS and was managed with IV and oral anticonvulsants and advised for the behavioral therapy to follow on out-patient basis. Neurosurgical consultation was also advised in case of intractable seizures for possible hemispherectomy, which has a success rate of 85% in selected cases. DDMS is a rare clinical entity and furthermore, status epilepticus is an unusual presenting complaint. The physicians must be aware of this relatively uncommon clinical presentation of status epilepticus in order for a timely and appropriate management and also for the outcome of a better long-term prognosis.

### **Speaker Biography**

Ghimire Pradesh has completed his MBBS from Manipal College of Medical Sciences affiliated to Kathmandu University, Nepal. He is a third year Radiology Resident in Bicol Medical Center affiliated to Bicol University, Philippines.

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