# SUBACUTE SCLEROSING PANENCEPHALITIS IN CHILDREN: A CASE SERIES

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ICNC 2022
17th INTERNATIONAL CHILD NEUROLOGY CONGRESS
ANTALYA, TURKEY | OCTOBER 3-7, 2022

## OBJECTIVES

Subacute sclerosing panencephalitis (SSPE) is a rare and progressive inflammatory disease of the central nervous system due to aberrant measles virus with a fatal outcome. In acute fulminant SSPE, the disease evolves rapidly leading to death within three months. In Indonesia (1985-1991), 12 cases of children with SSPE were reported with stage II clinical manifestation. Currently, there is no cure for SSPE, and eradication by effective vaccination program is considered to be more beneficial and cost-effective than any other high-level forms of control.

We aim to present three cases of SSPE in children at Dr. Cipto Mangunkusumo General Hospital, Jakarta, Indonesia.

## CONCLUSION

The occurrence of SSPE may be suspected in children with atonic seizure, motor regression, and jerking movements that are unresponsive to standard treatments, with previous history of measles infection. Patients with suspected SSPE should be monitored closely in the long term due to the high chance of fatal and progressive neurologic deterioration. Isoprinosine, intravenous immunoglobulin, and ketogenic diet may show benefits for patient with SSPE, however there is no existing consensus regarding the treatment of SSPE.

Scan here to see the criteria we used

## CASE SERIES

We report three cases of SSPE based on Dyken's criteria at different stages (Jabbour's clinical stages). The first case (patient A) is a-6-year old female with atonic seizure and spasticity as initial symptoms (clinical stage IIIB), the second case (patient B) is a-6-year old male with motor regression and frequent spasms (clinical stage IIIA), and the third case (patient C) is a-6-year old male with gait abnormality, jerking movements, and inability to walk (clinical stage IIIA). Risk factors for SSPE in these patients include male sex, low socioeconomic class, and history of measles infection at a younger age. Brain imaging of these patients shows no characteristic findings for neurometabolic disease or SSPE. The typical characteristics of electroencephalography (EEG) were found in one patient (C) along with elevated cerebrospinal fluid (CSF) and serum measles immunoglobulin G (IgG), leading to the diagnosis of SSPE. All patients received Isoprinosine (50 mg/kgBW/day) and supportive treatments. Treatment with a ketogenic diet or intravenous immunoglobulin was also attempted when possible, and all patients are currently on long-term monitoring and follow-up.

| Measles IgG test | Normal values IU/mL | Unit | Patient | | |
|---|---|---|---|---|---|
| | | | A | B | C |
| Serum | Negative <0.2<br>Equivocal ≥0.2 - <0.275<br>Positive ≥0.275 | IU/mL | 6.63 | 6.90 | 8.79 |
| CSF | | IU/mL | 6.75 | 6.50 | 8.70 |

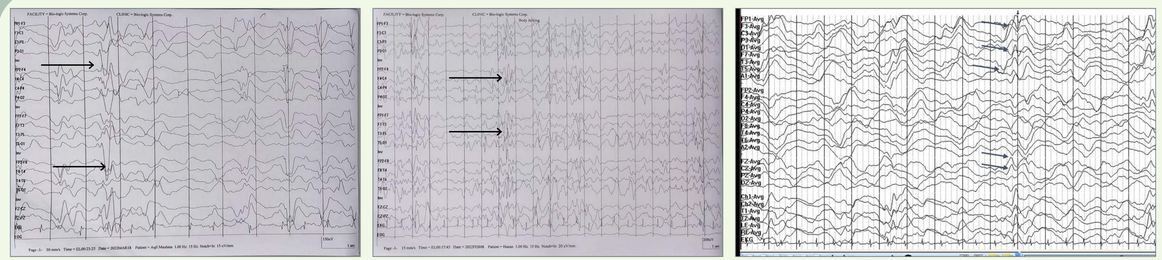

Fig 2. Electroencephalography of patient A, B, and C
Patient A and B : Generalized hypofunction and epileptiform discharges.
Patient C: Lateralized periodic discharges at left frontotemporal with generalized hypofunction.

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## ACKNOWLEGDEMENT

We would like to appreciate International Child Neurology Association for ICNC Bursary/Travel Award 2022 which supported the presenting author to attend 17th ICNC 2022.