the same disease was obtained in 2 cases (50% of NF1 and 100% of IP).

**Conclusions** The leading NCS in this study was SWS. 70% of children with NCS presented with seizures. The commonest type of seizure among them is GTCS. Children with TSC had seizures refractory to anticonvulsants. Careful evaluation of NCS children can reveal problems like Learning disabilities.

**Recommendation** All children with neurocutaneous markers should be evaluated and investigated in detail to detect neurological affection.

**Keywords** Neurocutaneous syndromes, Hypomelanosis of Ito, Sturge Weber Syndrome, Tuberous Sclerosis Complex.

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**PO-0819** CASE SERIES OF CHILDREN WITH NEUROCUTANEOUS SYNDROMES

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**Introduction** Neurocutaneous syndromes (NCS) are a heterogeneous group of disorders characterised by abnormalities of both the integument and central nervous system that are believed to originate from a defect in differentiation of the primitive ectoderm. Cutaneous manifestations usually appear early in life and progress with time, but neurological features generally present at a later age.

**Aim** To study the clinical profile of children with neurocutaneous syndromes and their various symptomatology, the seizure types and the response to treatment.

**Subjects and methods** A retrospective cross-sectional study was conducted in the Department of Paediatrics, Pushpagiri Medical College Hospital, Tiruvalla, during the period from January 2013 to June 2013. Children between the age group 0 and 15 years were included in the study based on the standard diagnostic criteria for different NCS. Investigations done were CT, MRI, EEG, and skin biopsy for appropriate cases.

**Results** The study population comprised of 10 children (5 boys, 5 girls). The various forms of NCS observed were Sturge Weber syndrome (SWS) – 4 Neurofibromatosis (NF1)- 2, Hypomelanosis of Ito (HOI) – 2, Tuberous sclerosis complex (TSC) – 1, and Incontinentia pigmenti (IP) – 1. A total of 8 children (80%) presented with neurological symptoms and the remaining 2 (20%) presented with cutaneous symptoms of which 1 was found to have learning disability on evaluation. The neurological problems were,70% had seizures of which100% were SWS and TSC, 50% were HOI and NF1. 72% had generalised tonic clonic seizures (GTCS) and 28% had focal seizures. The child with TSC showed refractory epilepsy. Developmental delay was detected in 50% of cases and maximum delay was seen in HOI. Family history of the same disease was obtained in 2 cases (50% of NF1 and 100% of IP).

**Conclusions** The leading NCS in this study was SWS. 70% of children with NCS presented with seizures. The commonest type of seizure among them is GTCS. Children with TSC had seizures refractory to anticonvulsants. Careful evaluation of NCS children can reveal problems like Learning disabilities.

**Recommendation** All children with neurocutaneous markers should be evaluated and investigated in detail to detect neurological affection.

**Keywords** Neurocutaneous syndromes, Hypomelanosis of Ito, Sturge Weber Syndrome, Tuberous Sclerosis Complex.