4×3cm diameter mass located in the inferior vermis and 4th ventricle advancing to foramen magnum.

Conclusion Swallowing difficulties in children is generally due to mild problems such as gastroesophageal reflux, esophagitis or food allergies. If swallowing difficulty is together with weight loss and is persistant for months intracranial pathologies, as in our case, should be searched.

543 INSIDE THE MIND OF AN ANGEL

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R Kyerematen. *Department of Paediatrics, Royal Cornwall Hospital, Truro, UK*

Background Angelman syndrome is a neurogenetic disorder characterised by severe learning difficulties and speech impairment, motor difficulties including jerky movements and ataxia, and seizures.

Epilepsy associated with Angelman syndrome consists of a variety of seizure types. Typically EEG shows a distinctive pattern which can aid diagnosis, but MRI scan shows no abnormality.

Although there is a known association between epilepsy and hippocampal sclerosis in the general population, the development of hippocampal sclerosis following a prolonged convulsion has not been described in a child with Angelman syndrome.

Methodology Literature search was carried out to review and compare similar reported cases. This elucidated that the association between hippocampal sclerosis and Angelman syndrome has rarely been cited.

Results The case presented here is a 3 year old girl with Angelman syndrome due to de novo micro deletion of chromosome 15, who suffered a prolonged convulsion and subsequently developed a persistent hemiplegia. Serial MRI scans demonstrate initially normal brain architecture and appearances, then the evolution from mild hippocampal swelling two days after the acute insult, to frank hippocampal sclerosis, as well as changes to the left cerebral hemisphere, several months later.

Conclusion This case evidences the development of hippocampal sclerosis following acute prolonged convulsion in a child with Angelman syndrome, and implicates this pathogenesis in the natural history of Angelman syndrome.

544 BENIGN EXTRA AXIAL COLLECTION OF INFANCY - A CASE REPORT

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A Kage. *Paediatrics, Kettering General Hospital, Kettering, UK*

Benign extra axial collection of infancy is a benign clinical entity characterized by rapid head enlargement in an infant with normal neurodevelopment.

We report on an infant who was referred at 3 months with rapidly increasing head circumference. The development was normal and there was a family history of macrocephaly. MRI brain showed normal ventricles with no hydrocephalus. There was significant prominence of subarachnoid space, particularly in fronto- parietal regions. Interestingly, there was moderate degree of cerebral atrophy.

Paediatricians should consider this diagnosis in any infant with rapid head enlargement and normal neurodevelopment. It is a benign condition that requires no surgical intervention because it often resolves spontaneously. The age of onset varies, but it is often seen in the first year of life, more often in boys, when an infant is noticed to have rapid head enlargement. It should not be confused with hydrocephalus or any other intracranial pathologies that are often associated with abnormal neurodevelopmental milestones. The persistence of the subarachnoid fluid collection beyond 2 years of age or a change in neurodevelopment calls for further evaluation to exclude intracranial pathology.

545 VICI SYNDROME ASSOCIATED WITH SENSORINEURAL HEARING LOSS AND LARYNGOMALASIA

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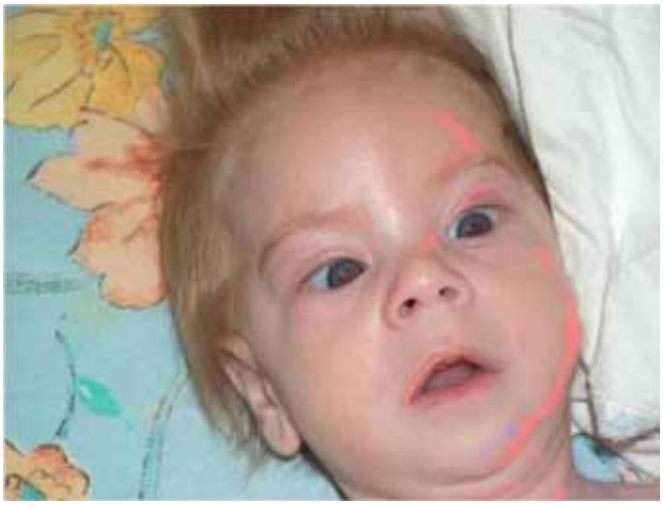
[1]M Özkale, [2]İ Erol, [3]A Gümüş, [1]Y Özkale, [1]B Kılıçaslan, [2]F Alehan. *[1]Department of Pediatrics; [2]Department of Pediatrics, Division of Child Neurology; [3]Department of Pediatrics, Division of Cardiology, Baskent University Hospital, Adana, Turkey*

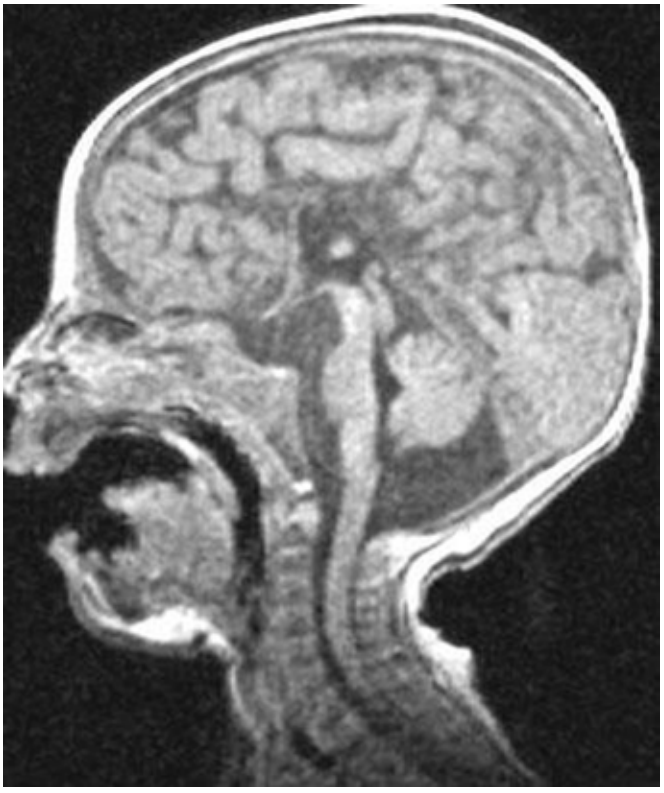
Background and Aims Vici syndrome is characterized by albinism, hypopigmentation, agenesis of the corpus callosum, catarats, immundeficeny, recurrent severe infections, hipertrophic cardiomyopathy and psychomotor retardation. To the best of our knowledge, this is the first reported case of a Turkish patient with Vici syndrome.

Case Report A 3 months-old girl admitted to our hospital for bronchopneumonia, stridor and failure to thrive. Physical examination revealed marked hypopigmentation of the skin with silvery hair, and dysmorphic features including highed-arched palate, micrognathia, generalize hypotonia, truncal ataxia with absense of deep-tendon reflexes (Figure 1). Ophthalmological examination revealed bilateral anterior subcapsular cataracs, and ocular albinism. Metabolic screening was normal. Magnetic resonance imaging of the brain showed agenesis of corpus callosum together with delayed myelinisation of cerebral white matter and hypoplasia of the cerebellar hemisphere and brainstem (Figure 2). Echocardiography was demonstrated hypertrophic cardiomyopathy. Odiological examination showed deafness on the left ears. Direct laryngoscopy was performed due to stridor and revealed laryngomalasia. She had immunological abnormalities including, decreased CD3+ (%38.1), CD3+/CD4+ (%31.1), CD3+/CD8+ (%7.2). Ceftriaxon and β-blocker were given for bronchopneumonia and cardiomyopathy. She had been lost due to broncopnomonia in an other hospital at the age of 6 months.

Conclusion Vici syndrome is considered in the different diagnosis of infants presenting with congenital agenesis of the corpus callosum. As until now 14 patients with Vici syndrome were reported we want to draw attention to this rare syndrome.



Abstract 545 Figure 1



Abstract 545 Figure 2

546 A GIANT ORBITAL PLEXIFORM NEUROFIBROMA WITH MASSIVE INTRACRANIAL EXTENTION IN A NEWBORN

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[1]N Yazici, [2]H Gulcan, [3]O Alkan, [4]R Yaycioglu, [5]N Bal, [1]F Sarialioglu. *[1]Department of Pediatric Oncology; [2]Department of Neonatology; [3]Department of Radiology; [4]Department of Eye Disorders; [5]Department of Pathology, Baskent University, Adana, Turkey*

Orbital masses in newborns are rare. Herein a newborn with a giant orbital tumor is presented. A 10-day female baby was admitted to hospital with proptosis. She was the first child of a 29 year-old mother, was born from an uneventful pregnancy. Her birth weight was 3000 gr. No consanguinity between the parents and history of neurofibromatosis in family were present. In physical examination, the baby had bilateral prominent proptosis and rest of the physical examination was unremarkable. The initial diagnosis was metastatic neuroblastoma. MRI of the brain showed a huge mass involving bilateral cavernous sinus, perimedullary cistern, orbita and orbital apex. Neuroblastoma markers including urine VMA, NSE, bone-marrow aspiration examination, abdominal ultrasonography and a two-diamentional chest x-ray were normal. Although the initial radiological diagnosis was plexiform neurofibroma, an open biopsy was performed to rule out other possibilities specially orbital malignant tumors. Histopathological diagnosis was a typical plexiform neurofibroma. The child has been treated with palliative measures and for seizures. Although it was a benign histology, the clinical picture was drastic. Like an infant with a malignant tumor chemotherapy was planned. But the family refused chemotherapy and any other form of antineoplastic therapy. In conclusion, the plexiform neurofibroma must be taken in consideration in newborn infant with orbital tumors.

547 CLINICAL PARTICULARITIES IN 2 CASES WITH POLYRADICULONEUROPATHY

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[1]SI Iurian, [2]S Iurian, [3]G Gradinariu, [3]A Vidrighin, [3]B Mehedintu. *[1]Research Department, Pediatric Clinic, Lucian Blaga University; [2]Clinical Laboratory, Pediatric Hospital; [3]Pediatric Clinic, Sibiu, Romania*

Background and Aims Guillain-Barré syndrome (GBS) is an acute polyradiculoneuropathy with weakness and diminished reflexes. The authors present clinical peculiarities in 2 cases diagnosed with GBS.

Methods The authors present 2 cases: a 18 month-old male admitted because of unstable walking (1st case) and severe lower extremities pain for the 2nd case (5 year-old boy).

Cases history: upper respiratory tract illness 3 weeks before symptoms onset; no vaccinations, surgical procedures or trauma prior to disease.

Clinical exam:

1. 1st case presented respiratory signs (dysphonia, slurred speech, short breath);
2. 2nd case was admitted for severe legs pain.

In addition, both cases were characterized by symmetrical extremities weakness, legs sensory changes (paresthesias, numbness), intense nuchal rigidity, positive Brudzinski sign, abolished *osteo-tendinous reflexes* in both upper/lower limbs and no abdominal reflexes. There were performed electromyography (EMG), nerve conduction velocity tests (NCT), serologic and cerebrospinal fluid (CSF) analysis.

Results CSF analysis identified albumino-cytologic dissociation: elevation of CSF protein with normal white blood cells count. The serologic studies shown normal titers for cytomegalovirus, Epstein-Barr virus and *Mycoplasma.* The NCT and EMG have proved severe demyelinating neuropathy and distal conduction block. Differential diagnosis: authors excluded meningitis, myopathies, poliomyelitis, polymyositis and myasthenic syndromes. The patients were treated with intravenous immunoglobulins with good clinical evolution.

Conclusions The authors presented 2 cases with GBS secondary to respiratory infections. Cases peculiarities.

1. Even though small children have the lowest risk, 1st case presented very early onset;
2. Both cases presented intense nuchal rigidity.

548 LINEAR GROWTH AND BODY MASS INDEX IN INFANTS AND CHILDREN AFTER CAUSTIC INGESTION

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[1]A Soliman, [2]W Saleem, [1]A Elawwa, [1]M Wagdy, [1]H Abdelrahman. *[1]Pediatrics; [2]Pediatric Emergency, Hamad Medical Center, Doha, Qatar*

Caustic injuries continue to be a significant morbidity in the pediatric patient group. Management of caustic ingestion in children remains a difficult challenge, with the outcome ranging from an asymptomatic state to esophageal strictures and variable effects on linear growth and weight gain.

We recorded and analyzed the growth data of 10 children ranging in age from 1 to 4 years with caustic ingestion presented from 2005 to 2007 and treated at Hamad Medical Center. Initial management consisted of prompt endoscopy and early institution of steroids and antibiotics. The decision on esophagoscopy was made on the basis of drooling and dysphagia. Significant esophageal burns were confirmed in all of them and subsequently five of them were managed successfully by repeated dilation due to multiple strictures.

None of the patients had underweight and/or stunting for 2 years after treatment. However, the BMI decreased from 16.77 +/– 3.5 kg/m2 to 16.26 +/– 2.9 kg/m2 and the height standard deviation score (HtSDS) decreased significantly from (-) 0.09 +/– 0.99 to (-) 0.58 +/– 1. Children with multiple strictures that required