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Introduction Congenital toxoplasmosis, a parasitic disease caused by the protozoon toxoplasma gondii is one of the classic intrauterine infections. The combination of hydrocephalus, retinochorioditis, intracranial calcification and parenchymal necrosis is rare. The majority of infected newborns present with uncharacteristic symptoms or remain asymptomatic. We report on a newborn with congenital toxoplasmosis and extensive cerebral findings.

Case Report Term male newborn, pregnancy and delivery uneventful, no maternal toxoplasmosis screening, admission on day 18 due to lethargy, sucking and muscular weakness, increase in head circumference of 4 cm since birth with gaping cranial sutures and recurrent cerebral convulsions. Brain ultrasound demonstrated distinctive ventriculomegaly with multiple intraventricular filaments. Liquor findings revealed elevated protein and positive toxoplasmosis PCR. Serologically toxoplasmosa antibodies showed positive as well. Treatment was commenced with pyrimethamine, sulfadiazine and folin acid. Ophthamological examination showed microphthalmus and retinal scars; NMR reveals multiple necrosis of basal ganglia and cerebellum. Multiple neurosurgical interventions were indispensible due to progressive intraventricular filament formation and subdural hygroma. Furthermore, severe thrombosis of the vena cava superior and the subclavian veins developed due to extensive clotting activation. Catheter intervention to achieve recanalization was performed with subsequent enoxaprarin therapy. After development of sinus thrombosis and progressive intracranial haemorrhage intensive care treatment was limited. The baby deceased after 28 days of treatment.

Discussion Our case demonstrates that though maternal screening is available and despite of existing treatment options severe courses of the disease are still possible. In case of excessive increase in head circumference an elaborate search for intrauterine infections should be mandatory.

540 PERINATAL (FETAL AND NEONATAL) DIAGNOSIS AND EVOLUTION OF CARDIAC TUMOURS

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Cardiac tumors are rarely symptomatic and highlighted in the fetus when the size and position do not interfere with intracardiac hemodynamic. Objectives. To present four cases of cardiac tumors, confirmed by Doppler echocardiography (Echo) performed in the first 14 days postnatal, 2 of which were already highlighted by fetal echocardiogtaphy. Cases presentation. Fetal echocardiography showed 3 and respetively 4 intracardiac mass, well circumscribed, oval, 6–12 mm diameter, with echogenic appearance increased from normal cardiac structure, located in the IVS and the posterior wall of the LV, slightly protrudes in the lumen but no significant obstruction of LV outflow tract. Postnatal Echo confirmed the fetal echocardiography diagnosis multiple cardiac rhabdomyama. ECG: no suggestive changes. Chest X-Ray: cardiomegaly. One of cases was later diagnosed with tuberous sclerosis Bourneville. Fetal echocardiography not extracardiac changes detected in this case. The three cases of rhabdomyoma evolved according to age, without major cardiac distress and while echocardiography showed mild involution of tumors size without complete disappearance In the fourth case, Echo in the neonatal period revealed atrioventricular septal defect with intracardiac masses, 2 of 3 pedicled, non obstructive, pleading for a multiple cardiac fibroma. Not cardiac arrhythmias were detected fetal and postnatal development. Conclusions. fetal ultrasound screening and especially at older age of pregnancy may reveal the presence of cardiac tumors, mainly rhabdomyoama, then confirmed by Echo postnatal. Monitoring these tumors both in utero and post natal to allow early detection of obstructive disorders, with sometimes severe cardiac distress and requiring cardiovascular surgery.

541 THE BIOMECHANICAL EVALUATION OF GAIT IN MONITORING OF TREATMENT IN CHILDREN WITH CEREBRAL PALSY-PRELIMINARY DATA

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The methods enable the objective gait evaluation of the gait in children with cerebral palsy (CP). One of the indexes used to the analysis is Gillette Gait Index (GGI), complying 16 clinically meaningful kinematic and three-dimensional parameters. The study was conducted with application of the three-dimensional system of gait analysis BTS Smart. Spatial-time parameters of gait and courses of angles of joint of lower limbs were determined on the basis of conducted research. Those parameters were used in estimation of Gillette Gait Index. The analyzed group consisted of 12 cerebral palsy children at the age of 5–13 years. All the children were assessed by the team before and after the botulin injection. The authors present the result of on the base of one of one patient, a girl at the age of 12 with right-sided spastic hemiparesis. The patient was evaluated three-times: before the botulin treatment, then three and six months after botulin injection. The value of the mean antetorsion of the pelvis in the saggital plain is now comparable to the healthy children at the same age. The GGI index improved in both lower extremities, for the right one within 24.14% and for the left one up to 40.69%. The authors regard the results presented above as the pilot-study; the evaluation of the larger groups of children with cerebral palsy is being planned. In the authors 'opinion the objective method of CP children gait evaluation may be the helpful tool for clinicians to optimize the way of CP children treatment.

542 A RARE CAUSE OF SWALLOWING DIFFICULTY THAT SHOULD BE ALWAYS IN MIND

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Aim Swallowing difficulty among children is not a frequent complaint. Generally this problem is associated with gastroesophageal reflux, esophagitis, and rarely is seen as an outcome of esophageal strictures. We report here a boy who was admitted to our clinic with swallowing difficulty but finally had a different diagnose.

Material and Method A 6-year-old boy was admitted due to swallowing difficulty. The family was in France and the child had this complaint almost for 4 months. The boy had lost 5kg but still was in the 50th centile both for weight and height. His physical, neurological and fundoscopic examinations were normal. But he was in a bad mood and seemed exhausted. He told that he could not swallow large particles and his portions became smaller gradually. Esophageal narrowing/stricture was thought thus Barium enema was performed and showed a normal esophagus. Then upper gastrointestinal endoscopy was performed, Endoscopical esophagitis and gastritis was confirmed by pathology with H. pylori gastritis. Although the boy was put on PPI and antibiotics his complaints did not improve, and he was still in a depressed mood. Finally a cranial MR was taken and showed a

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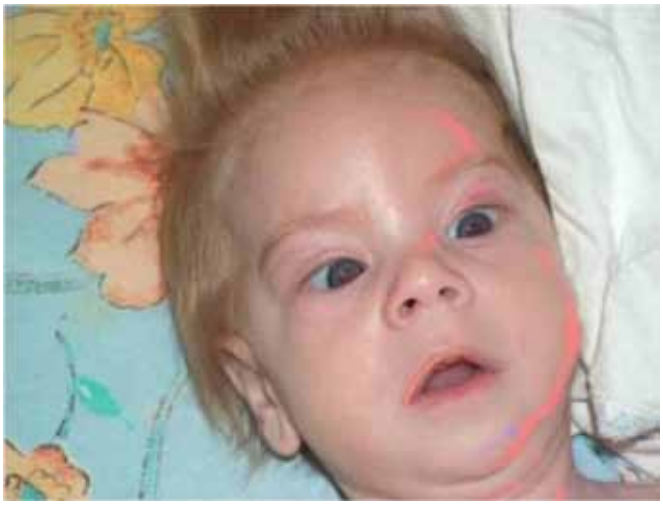
Background and Aims Vici syndrome is characterized by albinism, hypopigmentation, agenesis of the corpus callosum, catarats, immundeficeny, recurrent severe infections, hipertrophic cardiomyopathy and psychomotor retardation. To the best of our knowledge, this is the first reported case of a Turkish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