agreement with our assessment. Genetic testing was subsequently positive for congenital myotonic dystrophy. Parents finally consented to withdrawal of intensive care at day 64 of life and he died shortly after extubation.

**Conclusion** Critical care decision on withdrawal of intensive care can be a very traumatic experience for families. It is essential to follow the guidance available. As paediatricians we are advocates for the baby but at the same time we have to be empathetic and considerate to the sentiments of the family.

## 535 NECROTIZING ENTEROCOLITIS IN A NEWBORN FOLLOWING INTRAVENOUS IMMUNGLOBULIN TREATMENT FOR HEMOLYTIC DISEASE

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ABO iso-immunization is the most frequent hemolytic disease of the newborn. Treatment depends on the total serum bilirubin level, which may increase very rapidly in the first 48 h of life in cases of hemolytic disease of the newborn. Phototherapy and, in severe cases, exchange transfusion are used to prevent hyperbilirubinemia encephalopathy. Intravenous immunoglobulins are used to reduce exchange transfusion. Herein we present a female infant who was admitted to the our NICU because of ABO immune hemolytic disease and after two courses of 1gr/kg of IVIG infusion, she developed NEC. Administration of IVIG to newborns with significant hyperbilirubinemia due to ABO hemolytic disease should be cautiously employed and always administered under strict.

## 536 NEONATAL HSV ENCEPHALITIS: CONTROVERSIES OVER DIFFERENT THERAPEUTIC APPROACHES AND THEIR EFFECTS ON NEURO-DEVELOPMENTAL OUTCOMES

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**Background and Aims** Neonatal HSV encephalitis is well described and known to cause morbidity. However, there is no consensus regarding its optimal treatment, especially around using suppressive oral therapy after intravenous acyclovir. We aim to discuss treatment controversies and review possible neurodevelopmental outcomes in such cases.

**Methods** We report a case of vertically transmitted neonatal HSV-1 encephalitis and review existing literature on available treatment options (PubMed, EMBASE).

**Results** Our patient became pyrexial (39.0C) and lethargic on day 7 of life. Investigations revealed a raised CRP (80mg/l) and CSF pleocytosis (WCC-26/$mm^3$, 90% lymphocytes) with normal CSF biochemistry. IV antibiotics were empirically started. After developing encephalopathy and seizures on day 2 of illness, IV acyclovir was added. CSF PCR was positive for HSV-1. EEG showed multifocal irritability/excitability and asymmetrical temporal lobe activities. MRI showed low signal intensity on the ADC map in the medial temporal lobe cortices bilaterally and the right inferior frontal cortex.

21 days of IV acyclovir were completed, following which a repeat CSF sample was negative for HSV-1 PCR. IV antivirals were substituted with oral acyclovir at 1500mg/$m^2$/dose BD for twelve months.

**Conclusions** Literature review reveals controversies in treatment. Repeating HSV PCR at the end of IV treatment is not universally supported. Regarding suppressive oral acyclovir, some studies support doses of 1000–1740mg/$m^2$/dose BD while others favour a 300mg/$m^2$/dose TDS regime. Its optimal duration (6months, 12months or longer) is unclear. Neurodevelopmental outcomes mostly depend on the severity of the initial insult; Evidence that different suppressive treatments influence outcomes is poor.

## 537 AN IMPORTANT CAUSE OF DYSKINESIA

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Kernicterus has been referred to a disease of the past. However, we report two cases of kernicterus presenting with a dyskinetic movement disorder. Both cases had neonatal jaundice and were well until the age of 3 years with normal intellect. On examination dystonia, dyskinesia and chorea were seen. Further examination revealed an upgaze palsy and auditory neuropathy.

Kernicterus describes a neurological syndrome resulting from deposition of unconjuated bilirubin in basal ganglia & brainstem nuclei.

With the recent NICE guidance for jaundice therapy these cases highlight the importance of rigorous treatment of hyperbilirubinaemia. They also remind us to consider kernicterus as a diagnosis in a child presenting with a movement disorder and normal intellect.

## 538 THE VALUE OF NEAR-INFRARED SPECTROSCOPY (NIRS) IN PERINATAL ASPHYXIA-A CASE REPORT

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**Background** Perinatal asphyxia remains a challenging entity. NIRS offers a method to continuously monitor cerebral oxygen saturation.

**Aim** To obtain insight into haemodynamic changes during hypothermia and rewarming in perinatal asphyxia using NIRS.

**Methods** We report of an asphyxiated patient (37+6 weeks', Apgar 6 and 8 at 1 and 5 minutes, first arterial blood gas pH of 6.67, base deficit –25). NIRS was started during the first hour of life and continued for a total recording time of 125 hours. Simultaneously, we measured brain function using amplitude-integrated electroencephalography (aEEG). On day 7 magnetic resonance imaging (MRI) has been performed. After discharge, the patient was reassessed neurologically.

**Results** The initial cerebral rSO2 was 65%. When cooling was started FTOE was 0.28. At 33.5°C FTOE had decreased to 0.20, cerebral rSO2 increased to 70%. After rewarming cerebral rSO2 was 85%, and FTOE 0.11. Initially, aEEG showed a mixed burst-suppression and discontinuous pattern which improved to a discontinuous pattern only during the first 12 hours. After rewarming aEEG normalized and showed developing sleep-wake cycles. MRI did not show any signs of hypoxic damage. After discharge the patient presented neurodevelopmentally normal.

**Conclusion** After having cooled down the patient, both NIRS and aEEG showed an improvement (increase of rSO2, decrease of FTOE, loss of burst-supression in aEEG). aEEG displays cerebral function, cerebral NIRS expands information to cerebral oxygen supply and extraction. MRI and neurodevelopmental assessment proved the observed aEEG and NIRS data.

## 539 SEVERE FORM OF CONGENITAL TOXOPLASMOSIS WITH EXTENSIVE CEREBRAL FINDINGS

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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