

**P504 SANJAD-SAKATI-RICHARDSON-KIRK SYNDROME**Aamir Al Mosawi\*. *Baghdad Medical City, Baghdad, Iraq*

10.1136/archdischild-2019-epa.840

Early diagnosis of rare genetic disorders plays a vital role in preventing the disorder through appropriate genetic counseling. Sometimes early recognition of a rare disorder can help in reducing the associated morbidity and mortality.

The syndrome of severe growth retardation, mental retardation, and chronic hypocalcemia caused by hypoparathyroidism was first reported by Sanjad, Sakati, and Abu-Osba in 1988.

Sanjad, Sakati, and Abu-Osba presented a part of a full description of the syndrome in five infants at the 58<sup>th</sup> Annual Meeting of the Society for Pediatric Research, Washington DC, May 1988. Later, they published a more completed description of the syndrome in 1991. The paper of 1991 which was authored by more authors, and included twelve patients.

However, the syndrome was first fully described in 1990 by Ricky J Richardson from the Sick Children Hospital of Great Ormond Street in London, and Jeremy MW Kirk from St Bartholomew's Hospital in London.

Richardson and Kirk emphasized that this association of a previously undescribed congenital anomalies represented a new syndrome that was observed in eight children of middle eastern origin.

They thought that early recognition of this rare disorder may decrease the associated morbidity and mortality.

The total number of the reported patients with Sanjad-Sakati-Richardson-Kirk syndrome is 103.

Sanjad-Sakati-Richardson-Kirk syndrome has not been reported before in Iraq. The main aim of this paper is to report the first case of this syndrome in Iraq which is the case number 104 in the world.

Before referral, the child didn't receive management that can be described as appropriate.

The hypocalcemic seizure, he had was treated with low dose vitamin D followed with low dose of one alpha hydroxycholecalciferol (2 drops daily), and the boy continued to have seizures.

The calcium level remained below 7 mg/dL, but it was ignored by the treating physicians, and electroencephalography was performed and showed frequent focal epileptic discharges more prominent in central leads and mild slowing of cerebral activity.

The child was treated with anticonvulsant medications mostly sodium valproate which reduced the seizures, but didn't stop.

After referral, the child's hypocalcemia was treated with the appropriate doses of one alpha hydroxycholecalciferol. Calcium level was maintained above 8 mg/dL. Seizures stopped and sodium valproate was stopped.

**P505 BRAIN ABSCESS AS A FIRST MANIFESTATION OF ATRIAL SEPTAL DEFECT**Matija Bakoš\*, Filip Rubić, Dalibor Šarić, Daniel Dilber, Ivan Malčić, Dorotea Bartoniček. *University Hospital Center, Zagreb, Croatia*

10.1136/archdischild-2019-epa.841

A 6-year-old boy was admitted to our hospital due to a routine cardiac surgery of primum atrial septal defect (ASD) with cleft mitral valve. His medical history started seven months before, after he was admitted into the local hospital because of fever with chills, headache, nausea, stiff neck and vomiting. Diagnosis of meningitis was made, and antimicrobial treatment was initiated. Five days after, left side hemiparesis was detected. MSCT was performed and showed multiple brain abscesses on right part of basal ganglia and on the left along the temporal horn of ventricle system with a "mass effect". Furthermore, the threatening brain herniation was noticed. On that same day, he was transferred into our hospital for brain abscess drainage. Echocardiography was performed because of a still heart murmur and the diagnosis of primum ASD with cleft mitral valve was discovered. An indication for operative cardiac surgery after neurosurgical treatment was set. Control brain MR was been performed two weeks. On imaging there was no improvement, but muscular strength and general condition were better and the second drainage of brain abscesses with stereotactic method was performed. Streptococcus intermedius and mixed anaerobes were isolated from intraoperative smear. Antimicrobial treatment was in total of 6 weeks. Post-operative MSCT scan showed a significant regression of brain abscesses. Dental treatment for caries was done.

Six months later he went through an open heart surgery where an ASD primum was closed with pericardial patch and mitral valve plastic was done. Postoperative period was uneventful.

**Conclusion** Brain abscesses are rare, but serious, life-threatening neurological entities. The abscesses are usually deep seated and thin walled. Right parietal lobe is often more affected because of the direct blood flow to this area. Cyanotic congenital heart diseases are rare cause of brain abscess in children; however, an ECHO should be done as routine examination to exclude one.

**P506 THE PATTERN OF PRESENTATION IN CHILDREN WITH HERPES SIMPLEX ENCEPHALITIS**<sup>1</sup>Jennifer Cox\*, <sup>1</sup>Elizabeth Murphy, <sup>1</sup>Laura Whitla, <sup>1,2</sup>David Coghlan, <sup>1,2</sup>Montaseur Nadeem. <sup>1</sup>Tallaght University Hospital, Dublin, Ireland; <sup>2</sup>Trinity College, Dublin, Ireland

10.1136/archdischild-2019-epa.842

**Introduction** HSE is estimated to occur in approximately 1 in 250,000 to 1 in 500,000 individuals per year, with approximately one third of cases occurring in children and adolescents. Poorer prognosis and neurological sequelae are associated with delayed treatment of HSE.

We retrospectively reviewed all cases of HSV meningoencephalitis admitted to our centre over a ten year period from Jan 2008 to Jan 2018. Four cases were identified based on PCR confirmation of HSV 1 or 2 in cerebrospinal fluid samples. Here we discuss the clinical presentation, as well as electroencephalogram, neuroimaging and clinical outcomes, to highlight the importance of early recognition and timely treatment of such cases.

**Case series Case 1:** Nineteen month old male infant presented with prolonged focal febrile seizure, on a preceding history of pyrexia and lethargy for one day, and cough for one week. EEG showed excess slowing over both posterior regions, more marked on the left posterior region, consistent with bilateral cerebral dysfunction without epileptiform discharges. MRI

brain demonstrated deep white matter changes in the left parietal and posterior parietal/occipital regions and adjacent grey matter and two small punctate haemorrhages in the left mid-parietal region.

Case 2: Four week old male infant presented with recurrent focal seizures. No pyrexia or preceding symptoms were documented. EEG recorded two seizures with origin near the right midline with spread to right motor cortex and right hemisphere. MRI brain showed right deep white matter changes and left subcortical lesion.

Case 3: Eleven week old female infant admitted with pyrexia and lethargy. She had 6 episodes of febrile generalised tonic seizures over 48 hours, each less than 5 minutes in duration. EEG demonstrated some asymmetry and excess slow activities over right temporal region in sleep, with no epileptiform features. MRI brain was normal.

Case 4: Thirteen month old female presented with febrile status epilepticus, on a background history of pyrexia and diarrhoea for 3 days. EEG showed three epileptic spasms with background continuous epileptiform discharges, which were multifocal; predominantly right posterior and some frontal discharges were seen in sleep. MRI brain showed subtle high signal white matter changes in the frontal lobes bilaterally and multiple high signal areas throughout corpus callosum.

**Discussion** This case series demonstrates that HSV encephalitis should be considered in children with atypical febrile seizures which are prolonged, focal or recurrent. Timely recognition of HSE is essential to minimise patient morbidity and mortality.

**Interventions** We administered AURASTOP<sup>®</sup> at the dose of 1 teaspoon 2 times per day for 2–3 months and when an attack occurs, in children above 4 years of age, at a dose of 1 pouch.

**Main outcome measures** It has been used two validated questionnaire: Migraine Disability Assessment (MIDAS) and The six-item Headache Impact Test (HIT-6). MIDAS is a brief questionnaire and measures headache-related disability. HIT-6 was designed to provide a global measure of adverse headache impact. Treatment with AURASTOP<sup>®</sup> has been conducted for 3 months, with two sachets-day. Compliance of patients was monitored weekly by means of a weekly exchange of emails or phone calls to parents.

**Results** A reduction in MIDAS scores was observed comparing pre-treatment ( $46.48 \pm 8.35$ ) and post-treatment ( $9.78 \pm 18.16$ ) data. After the treatment 1/42 patients (2.38%) had worsened, 9/42 patients (21.43%) had not improved, 10/42 (23.91%) had gone up 1 class, 12/10 patients (28.57%) had gone up 2 classes, 10/42 (23.91%) had gone up 3 classes of MIDAS score. Therefore, the improvement of disability related to the headache is statically significant (Fisher's exact test). The reduction of HIT-6 score post-treatment ( $46.48 \pm 8.35$ ) compared with pre-treatment ( $62.55 \pm 5.50$ ) was statistically significant ( $P < 0.05$ ).

**Conclusions** AURASTOP<sup>®</sup> can be used as a prophylactic treatment and as symptomatic treatment in paediatric patients with headache.

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#### TANACETUM PARTHENIUM, GRIFFONIA SIMPLICIFOLIA AND MAGNESIUM AS SYMPTOMATIC AND PROPHYLACTIC TREATMENT FOR HEADACHE IN PAEDIATRIC PATIENTS

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10.1136/archdischild-2019-epa.843

**Objective** The aim of the this study is to evaluate the use and the self-perceived efficacy and tolerability of three nutraceutical components - Tanacetum partheninum, Griffonia simplicifolia and Magnesium - in children and adolescents with primary headaches without other comorbidities.

**Design** This is a longitudinal cohort study.

**Setting** The study was conducted as a prospective study. After the first examination and the prescription of exams, patients were subjected to one month baseline period without treatment to verify the number and the intensity of attacks.

**Patients** We enrolled a total of 42 children, 25 female (59,52%) and 17 male (40,48%). The average age of children at the time of recruitment is  $10,59 \pm 3,18$  years. The inclusion criteria employed have been: (a) aged between 3–17 years; (b) diagnosis of primary headache and/or migraine (including migraine with or without aura; chronic headache; persistent headache); (c) headache/migraine present for at least three months; (d) at least three migraine attacks per month in the last three months. The exclusion criteria have been: (a) patients with chronic pain of different nature; (b) secondary headache.

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#### PARAECHOVIRUS MENINGOENCEPHALITIS IN INFANCY; A CASE REPORT

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10.1136/archdischild-2019-epa.844

**Aims** This article aims to describe and highlight human paraechovirus (HPeV) as a cause of meningoencephalitis.

**Methods** A five week old (previously well) term male infant presented with a one day history of pyrexia and irritability. On examination, he was tachycardic, febrile and inconsolable. Baseline investigations showed serum white cell count (WCC)  $4.5 \times 10^9/L$  (46% neutrophils, 37% lymphocytes), CRP 11 mg/L. Cerebrospinal fluid (CSF) analysis showed WCC 72/cm, 43/cm and 114/cm in three samples (with 18% polymorphs and 82% mononuclear cells), protein 0.78 g/L, and glucose 3.3 mmol/L. Treatment with triple antibiotics and acyclovir was commenced pending further results. EEG on day 2 of admission was abnormal with low amplitude throughout and slow rhythmic activities. Due to ongoing severe irritability and persistent tachycardia, further investigations including an MRI brain and echocardiogram (ECHO) were also undertaken and were reassuringly normal. CSF and blood cultures were negative at 48 hours. However CSF polymerase chain reaction (PCR) result detected paraechovirus RNA; the CT value (crossing target) was 31.6 suggesting a relatively small viral load. CSF sampling was repeated 48 hours later and the WCC had improved to 15/54/38 cell/cm. CSF PCR was again positive for paraechovirus and the CT value had increased to 39.2 suggesting a decrease in the viral load. This correlated with the slow clinical improvement. Of note the EEG on day 4 of admission was again very abnormal, as was the follow up EEG 6 weeks later which showed some excess slowing