British Society for the History of Paediatrics and Child Health

1810 WHEN WAS MULTIAGENCY WORKING TO SAFEGUARD CHILDREN INVENTED?

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Background I was a Consultant Paediatrician and Designated Doctor for Safeguarding Children for Sheffield. I have completed an MA in the history of the interaction of health and education in Sheffield schoolchildren, 1870–1906. I am currently undertaking a PhD in Safeguarding Children in Sheffield, a rapidly expanding industrial city from 1870–1914.

Objectives To investigate whether there was evidence of multi-agency work in Sheffield from 1870–1900 to safeguard children from maltreatment.

Methods The study is based on prosecutions for child maltreatment as reported in local newspapers from 1870–1900, 19 years before and 11 years after the 1889 Prevention of Cruelty to, and the Protection of Children Act, and the setting up of an NSPCC office in the city.

Results 113 cases of child maltreatment by parents or step-parents, involving 260 children were found, 41 in the 19 years 1870–1889 (Group 1) and 72 in the 10 years 1890–1900 (Group 2). Group 1 involved mainly cases of non-accidental injury of a single child with only the presenting physical symptom, eg bruising, investigated by a single agency. In Group 2, whole families were holistically assessed by a range of professionals led by the local NSPCC inspector. These include doctors, police, sanitary inspectors, workhouse staff and education staff and cases of neglect predominated in this group.

Conclusions 1. The number of prosecutions trebled after the Children Act of 1889, as did the number and proportion of cases of neglect.
2. After 1889, the assessments were wide-ranging and holistic carried out mainly on working class children and families by middle class professionals and covered the child, siblings, home and parenting.
3. This study shows that multiagency assessments to Safeguard children were developed in the late nineteenth century England, a century before the government Every Child Matters and Working Together to Safeguard Children guidance.

British Paediatric Neurology Association

1812 A CASE REPORT OF EVOLVING NEUROMYELITIS OPTICA PRESENTING AS SYNDROME OF ANTIDIURETIC HORMONE SECRETION (SIADH) IN AN EIGHT YEAR OLD

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Background Optic neuritis is a rare demyelinating disorder, which involves the optic nerve. It can be a monophasic self-limiting illness following infection or vaccination. It can also be an initial presentation of a relapsing demyelinating disorder like multiple sclerosis or neuromyelitis optica spectrum of disorder (NMSOD). It characteristically affects aquaporin 4 receptor rich areas in the brain like hypothalamus, periventricular areas as well as optic nerve and spinal cord. This may lead to dysfunction in the hypothalamo-pituitary axis, hence syndrome of inappropriate antidiuretic hormone secretion (SIADH). This may manifest as hyponatraemia and when severe presents with confusion, agitation and convulsions.

We present a 8year old girl, presented with acute symmetric hyponatraemia due to SIADH. She had confusion, agitation with 2 brief generalized convulsions. Her initial S. Na was 120 mmol/l, serum osmolality 256mmol/l, urinary osmolality 356mmol/l, urinary sodium 55mmol/l, hematocrit is normal and serum albumin is 45mg/dl. Her sodium corrected and her symptoms disappeared.

Within 12 hours, she developed acute loss of vision. Her eye movements were painful. Color vision was completely absent in both eyes (0/14). Bilateral pupillary reaction had relative afferent pupillary defect. Her bilateral visual acuity was 6/9.

Her bilateral visual evoked potential (VEP) was delayed.

MRI brain, spine and optic tracts without gadolinium enhancement were inconclusive. The metabolic work up, including CSF studies were normal. CSF oligoclonal bands were negative. Anti-aquaporin-4 antibody and anti-myelin oligodendrocyte antibody were unavailable. She had a dramatic response to corticosteroids. But after 8 weeks her VEP is still prolonged in left eye.

Her disease progression is yet to be manifested, whether it is monophasic or a first episode of a multiphasic demyelination disease.

This case reports highlights the rare association of evolving NMSOD with SIADH, and the possibility of initial presentation as acute symptomatic hyponatraemia.

1813 PROGRESSIVE DYSTONIA AND NEURODEGENERATION WITH EYE OF THE TIGER SIGN IN MRI BRAIN – HALLERVORDEN- SPATZ SYNDROME

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Background Hallervorden- Spatz neurodegeneration is a rare neurodegenerative disorder. It is autosomal recessive. It is a familial brain degeneration characterized by cerebral iron deposition, mainly in globus pallidus.

It is characterized by progressive extrapyramidal and corticospinal degeneration and dementia. The phenotypic spectrum would be either classical which has an early childhood onset or atypical which has a late onset of disease (>10years).

The clinical course is characterized by extrapyramidal signs like dystonia, dysarthria and parkinsonism and corticospinal tracts involvement signs like spasticity, hyperreflexia and extensor toe sign. Pigmentary retinopathy, low intelligence, developmental delay and psychiatric disturbances are also documented.
Brain MRI has a distinct feature with eye-of-the-tiger sign characterized by bilateral hypointensity of the globus pallidus with a central area of hyperintensity on T2-weighted images. Peripheral blood acanthocytosis and low plasma pre beta lipoprotein are supportive evidence. Establishing PANK 2 variant is confirmatory.

Management is mainly supportive. Dystonia can be controlled with benzodiazepines, anticholinergics and botulinum toxin. Spasticity can be revealed with baclofen. Stereotactic surgical modalities such as thalamotomy, pallidotomy and deep brain stimulation of the globus pallidus can help to control symptoms.

We present a 5 year old boy who presented with tiptoe walking who gradually deteriorated neurodevelopmentally. He had declined in academic performance and progressive dystonia, dysarthria and spasticity was there. He had hyperreflexia but parkinsonism, chorea or myoclonus was not evident. The neuro-ophthalmologic examination was normal. Laboratory work up was normal including serum copper, ceruloplasmin levels. Blood smear had no acanthocytes. T2 weighted brain MRI was consistent with ‘eye-of-the-tiger’ sign. Genetic testing was not performed due to the cost effect.

The child is under multidisciplinary rehabilitation to improve quality of life.

This case report highlights the importance of considering this entity in progressive dystonia with neurodegeneration.

**Background**

Pediatric demyelinating disorders (PDDs) are rare neurological conditions reported among children. It has variable neurological presentations which challenge in diagnosis.

**Objectives**

This study was conducted to assess the clinical characteristics and outcomes of the PDDs reported to paediatric neurology unit, Teaching Hospital Karapitiya (THK).

**Methods**

A cross sectional study was conducted among neurological symptomatic patients attended, paediatric neurology unit at THK for five years duration. Data was extracted from clinical records available at paediatric neurology unit. All patients reported during study period were included.

**Results**

A total sample of 9245 children with neurological symptoms were reported during the study period. Among them, 20 children were diagnosed to have PDDs with 0.21% of five years period prevalence. Acute Demyelinating Encephalomyelitis (ADEM) (n=14, 70.0%) & Optic neuritis (n=4, 20.0%) were identified as common PDDs while Transverse myelitis and Neuromyelitis Optica spectrum of disorders (n=1, 10.0% for both) were identified as less common. No patient was identified with Multiple sclerosis. Half of the children with PDDs were male (n=10). Mean (SD) age of the presentation of PDDS was 13.1(3.0) years. Altered consciousness (n=12, 85.7%), headache (n=10, 71.4%) sensory deficits (n=8, 57.2%), convulsions (n=6, 42.8%) and acute psychiatric symptoms (n=5, 35.7%) were identified as common presentation for ADEM. All the children with Optic neuritis presented with reduced visual acuity. Blood investigations, viral isolation and neuroimaging were non-significant in all the children except who were diagnosed with ADEM. Majority of the children completely recovered following treatment (85.7%, n=12 for ADEM and 75.0%, n=3 for Optic neuritis). However, one death and one residual defect was reported among ADEM patients while urinary and bowel incontinence was reported for Transverse myelitis.

**Conclusions**

PDDs were rare among neurological symptomatic patients attended to pediatric neurology unit at THK. Clinical presentation, investigations, neuroimaging findings varied among them challenging the diagnosis. Majority had better outcome following treatment.