

1812 OUTCOME OF CHILDHOOD SYSTEMIC LUPUS ERYTHEMATOSUS (SLE) WITH LUPUS NEPHRITIS (LN)

doi:10.1136/archdischild-2012-302724.1812

¹M Ilyas, ¹A Salahuddin, ²RC Butterfield, ¹A Tolaymat. ¹Pediatrics; ²Center for Health Equity and Quality Research, University of Florida, Jacksonville, FL, USA

A total of 46 patients with SLE-LN were studied (91% females; mean age 13.0 years; mean follow-up duration 5.9 years). Initial renal biopsy showed that out of 46 patients, 2 (4.3%) had Class I, 14 (30.4%) had Class II, 8 (17.4%) had Class III, 20 (43.3%) had Class IV and 2 (4.3%) had Class V lupus nephritis. Based on the renal histopathology and clinical presentation, 23 (50%) patients received intermittent intravenous cyclophosphamide bolus with prednisone and Azathioprine (AZT) or Mycophenolate Mofetil (MMF). The remaining 23 (50%) patients were treated with corticosteroids alone or in combination with AZT or MMF.

Follow up renal biopsies were performed on 21 patients, 4 patients showed no change in histology, 8 patients showed histological improvement of lupus nephropathy, 9 patients showed progression of lupus nephropathy and 7 of these who progressed, were started on intermittent IV CYC bolus.

The clinical follow up revealed that out of 46 patients, 21.7% patients went into complete remission, 58.6% patients remained under control with immunosuppressant medications, 10.8% patients had clinically active disease with normal renal function and 10.8% patients had adverse outcome. The adverse outcome included one patient developed chronic renal insufficiency, three (3) progressed to end stage renal diseases and one died. Five-year kidney survival was 93.5% and patient survival was 97.8%.

Although IV CYC treatment has improved the mortality and morbidity in lupus nephritis but severe adverse effect makes it less than optimal for long term therapy.

1813 SERVICE PROVISION FOR CHILDREN WITH JUVENILE IDIOPATHIC ARTHRITIS (JIA) IN THE EAST OF ENGLAND (EOE); A COMPARISON WITH NATIONAL CENTRES

doi:10.1136/archdischild-2012-302724.1813

¹PJ Bale, ²K Armon, ³A Kavirayana. ¹Paediatric Department, Ipswich Hospital NHS Trust, Ipswich; ²Jenny Lind Children's Hospital, Norfolk and Norwich University Hospital, Norwich; ³Paediatric Rheumatology Department, Bristol Royal Hospital for Children, Bristol, UK

Introduction The EoE has no regional centre or clinical network for paediatric rheumatology. JIA has a prevalence of 1:100 (2), which suggests an estimated 1,200 cases managed in the 17 hospitals of the EoE. What sort of service do these children get, compared with agreed standards, and that provided by national and 'grid training' tertiary centres?

Aim To assess the service provision for children with JIA in the EoE against recommended standards of care as set out by ARMA/BSPAR 2010(1), and compare our results to national data.

Methods A questionnaire was sent to the Paediatric Rheumatology leads of the 17 hospitals in the EoE, focused on the service provision for paediatric rheumatology patients, against BSPAR/ARMA standards. Follow-up phone calls were made where necessary to ensure accuracy. Data from 15 National centres, including all 8 UK grid training centres were used for comparison (3).

Results 16 of 17 hospitals responded in the East of England.

Table 1. Shows the percentage of hospitals achieving each of the audited ARMA/BSPAR standards in EoE, compared with national centres, and grid training tertiary centres data alone. In summary, in grid tertiary centres 17 of 21 standards audited are achieved to a good standard (>85%), whereas in the EoE only 3.

Abstract 1813 Table 1

Standard	EOE (n=16)	National centres (n=15)	Grid Training (n=8)
New patient appointment >45 min	0.0	26.7	62.5
Follow up appointment >20 min	31.3	73.3	75
Paed rheum Nurse specialist	37.5	80.0	100
Paediatric Physiotherapist	87.5	86.7	100
Paediatric Occupational therapist	68.8	53.3	87.5
Access to Psychologist	56.3	80.0	87.5
Access to Ophthalmologist	100.0	100.0	100
Telephone helpline	43.8	73.3	100
Paediatric Anaesthetic list	50.0	73.3	100
Entonox available	37.5	73.3	100
Home Methotrexate training	56.3	66.7	87.5
Access to MTX in <4/52	100.0	93.3	87.5
Prescribe biologics	50.0	100.0	100
Recruit to Biologics registry	50.0	73.3	100
Have audited biologic use	25.0	46.7	75
Part of Clinical network	6.3	80.0	87.5
Recruit to research	25.0	60.0	87.5
Involved in undergraduate teaching	75.0	73.3	87.5
Child friendly clinic setting	81.3	93.3	100
Adolescent clinic	18.8	53.3	75
Referral to a specific adult rheumatologist	75.0	93.3	87.5

Conclusion The EoE hospitals fall seriously short in providing trained, specialist care for the estimated 1,200 children with JIA, suggesting serious inequality of access. Many standards are unachievable without commissioned resource. Our challenge is to improve provision within existing funding.

References

1. Arthritis and Musculoskeletal alliance (ARMA) Standards of Care for children and young people with Juvenile Idiopathic Arthritis 2010.
2. Children's Chronic arthritis association website. HYPER-LINK "http://www.ccaa.org.uk" http://www.ccaa.org.uk.
3. Dr Akhila Kavirayana (SpR Paediatric Rheumatologist) personal communication of data presented at BSPAR Sept 2011, as yet unpublished.

1814 PSYCHOPATHOLOGICAL PROFILE AND HEALTH RELATED QUALITY OF LIFE IN NARCOLEPSY WITH CATAPLEXY ACROSS CHILDHOOD AND ADOLESCENCE: A CASE-CONTROL STUDY

doi:10.1136/archdischild-2012-302724.1814

¹E Finotti, ²F Pizza, ³F Poli, ²C Franceschini, ³O Bruni, ²G Plazzi. ¹University of Bologna; ²Department of Neurology, University of Bologna, Bologna; ³Child Neuropsychiatry Unit, Sapienza University of Roma, Roma, Italy

Objectives The purpose of our study was to describe the behavioral aspects and quality of life of childhood narcolepsy with cataplexy (NC).

Methods We performed a case-control study based on self-administered questionnaires in 30 NC hypocretin-deficient patients, 39 epilepsy patients, and 39 healthy controls matched for sex and age.

Results Our population of children and adolescents with NC showed an increase in internalizing problems in line with previous reports, typically represented by withdrawal and depression symptoms, and somatic complaints. The two patients groups share higher scores than controls for anxiety disorders, attention, social and oppositional-defiant problems. Psychopathological profile in NC were found to be positively correlated with early NC onset,

diagnostic delay, nocturnal disturbed sleep, shorter sleep latency and greater number of Sleep Onset REM Periods at Multiple Sleep Latency Test. On the other hand, treatment and disease duration, positively influenced the behavioral evolution. The psychosocial health of pediatric NC also turned out to be worse than in healthy controls, while the physical health showed no significant differences.

Conclusions We found a specific psychopathological profile in a large pediatric NC sample, compared with another neurological chronic disease (epilepsy) and healthy controls. Symptoms of withdrawal, depression and somatic complaints, were specific of NC, and not observed in the two other groups. Effective treatment, and self-awareness of the disease should be promoted in NC children for the positive impact on behaviour and psychosocial health.

1815 SLEEP PROBLEMS AND ADHD. EPIDEMIOLOGICAL STUDY IN SCHOOL CHILDREN IN ANDALUSIA, SPAIN

doi:10.1136/archdischild-2012-302724.1815

¹A Lora-Espinosa, ²MJ Díaz-Aguilar, ³R Jiménez-Alés, ⁴N Palomino-Urda, ⁵R Hergueta-Lendínez, ⁶M Montes-Peña, ⁷J Murcia-García, ⁸N Martínez-Moral, ⁹J Barea-García, ¹⁰A Prosper-Gibert. ¹Servicio Sanitario Público Andalucía, Unidad Investigación Distrito Sanitario Málaga; ²Centro de Educación del Profesorado, Málaga; ³Servicio Sanitario Público Andalucía, Sevilla; ⁴Servicio Sanitario Público Andalucía, Granada; ⁵Servicio Sanitario Público Andalucía, Córdoba; ⁶Servicio Sanitario Público Andalucía, Jaén; ⁷Servicio Sanitario Público Andalucía, Cádiz, Spain

ADHD and sleep-problems frequently overlap and their relationship is complex and bidirectional. The association between ADHD and sleep-problems has been little studied in our community.

Objectives To find out the frequency of sleep-problems among ADHD children from 6–14 year old in Andalusia, Spain.

Methods Prevalence study. Target population: school-students 6–14y=686.332 children; centres=2.493. Multistage cluster sampling centres=74.

Method Questionnaires-DSM-IV-TR-ADHD and performance Vanderbilt-parents-and-teachers, sleep-questionnaire (BEARS), interview and medical examination.

Results N=1963.

Meet criteria-DSM-IV-R-ADHD=157, male=111, female=46; ADHD-IA=62, ADHD-HI=31, ADHD-C=64; control=197 Comorbidity ADHD-sleep-problems: Sleep-problems in the first year of life: 36%-ADHD, 25%-control. (p=0.03). Bedtime resistance:30%-ADHD, 6%-control.(p=0.000). Daytime sleepiness:10%-ADHD, 1.5%-control. (p=0.000). Night- Awakenings:14%-ADHD, 2%-control.(p=0.000). Snoring: 20%-ADHD, 5.6%-control.(p=0.000) Sleepwalking and sleep terrors:14%-ADHD, 2.5%-control.(p=0.000). Periodic limb movements in sleep: 51%-ADHD, 8%-control.(p=0.000). Enuresis:18%-ADHD, 4.6%-control.(p=0.000)Regular time for bed: only 36%-ADHD. Bedsharing:18%-ADHD, ≥ 3 sleep problems: 36%-ADHD. 12%- control. The association between ADHD subtypes and sleep-problems showed significant differences.

Conclusions The children with ADHD had more sleep-problems than control children.

The relationship between sleep disorders and ADHD should be considered by paediatricians as part of the global approach to the management of ADHD.

1816 SLEEP PATTERN AND SLEEP DISORDERS IN PRESCHOOL-AGED CHILDREN

doi:10.1136/archdischild-2012-302724.1816

M Taşar, H Boztepe, Ş Yıldırım, Y Dallar Bilge. *Pediatrics, Ministry of Health, Ankara Training and Education Hospital, Ankara, Turkey*

The purpose of this study determine sleep pattern, sleep disorders and factors affecting on preschool-aged children.

Material-method The research was conducted on 999 patient 2–6 years presenting patient out-patient clinic. Parents were given a survey containing questions about the sociodemographic and sleep.

Results 30.9% of children were entering to bed in the hours 20.00 to 22.00 and 67.0% of children were entering to bed in the hours 22.00 to 00.00. Sleeping and waking hours of children were found to be compatible with their parents (p=0.001). 50.2% of children with bedtime from 20.00 to 22.00 were fall asleep immediately and 22.6% of children with bedtime from 22.00 to 00.00 were fall asleep immediately (p=0.001). We found that 62.9% of children snoring, 72.5% of children with mouth breathing, 38.7% of the children stopped breathing, 79.3% of the children were restless sleep, 80.2% of children saw a nightmare, 43.1% of the children gnashed teeth during the sleep. We found that snoring, mouth breathing were more than and sleeping hours were later for son children and nightmares were more frequent in girls (p<0.05). We found that snoring, mouth breathing, restless sleep and frequent waking findings were more frequent in children with symptoms of attention deficit hyperactivity than in the other group.

Conclusions In the preschool-aged children, sleep disorders were common in. Sleep disorders were more frequent in children with hyperactivity symptoms. Primary care assessment of preschool-aged children should be questioned their sleep problems.

1817 DISCOVERED DEAD DURING SLEEP IN CAUSAL PATHWAY OF MORTALITY AMONG CHILDREN WITH CEREBRAL PALSY: CASE SERIES AND SYSTEMATIC REVIEW

doi:10.1136/archdischild-2012-302724.1817

¹L Holmes, ¹A Karatas, ¹D Kirk, ¹J Connor, ¹K Rogers, ¹F Miller. ¹Orthopedic Department, Nemours/A.I. Dupont Children Hospital, Wilmington; ²Biological Sciences, University of Delaware, Newark, DE, USA

Background and Aims To report thirteen cases of children with cerebral palsies (CP) discovered dead during sleep (DDDS) and to synthesize the research literature on CP survival.

Method We utilized case series to describe the common features among patients DDDS between 1993 and 2011. Using our medical records, we extracted data on demographics, treatment, and comorbidities. In addition using the appropriate search terms, we searched the Medline and other electronic data bases for articles published between 1966 and 2011. Further, we synthesized the literature and provided evidence on clinical conditions that could be associated with CP mortality in relation to DDDS, obstructive sleep apnea syndrome (OSAS) and sleep related breathing disorders (SRBD) management.

Results Between 1993 and 2011 there were 177 reported deaths, of which 13 occurred as a result of being DDDS at home. All the deceased patients had Gastrointestinal feeding tube, seizure, respiratory disorders, and were non-ambulatory. A greater proportion had scoliosis and hip dislocation surgeries. DDDS in our sample may be associated with these conditions; and OSAS/SRBD as evidenced in literature.

Conclusion Pulmonary problems and disorders were the most common co-morbidity, due probably to oxygen desaturation (< 70%), prolonged sleep apnea such OSAS and SRBD. Since there were no data on Polysomnography (PSG) and autopsy, it was difficult to account for specific sleep disorders that might have contributed to DDDS. We recommend a routine PSG and treatment of OSAS and SRBD, given their high prevalence among CP patients, especially those with disturbed nocturnal sleep and noisy breathing.

1818 SLEEP DISTURBANCES IN EPILEPTIC CHILDREN

doi:10.1136/archdischild-2012-302724.1818