

shortly after birth but milder variants present later. The phenotype in female heterozygotes is variable and depends on the pattern of X-chromosome inactivation in hepatocytes. This can range from severe neonatal encephalopathy to being completely asymptomatic throughout life. Survival and neurocognitive outcomes can be better than in severely affected males but are variable. Aggressive treatment of hyperammonaemic crises, dietary management with low protein diets, and alternative pathway therapy prevent further cognitive decline.

This case highlights the importance of early ammonia testing in encephalopathic patients regardless of age. A diagnosis of OTCD should be considered in patients with hyperammonaemia regardless of sex. Early recognition and appropriate treatment are critical to avoid severe brain damage and death.

G390(P) PICU OUTCOMES OF PAEDIATRIC ONCOLOGY PATIENTS IN A TERTIARY CENTRE

L Prasad, A Pickard, R Thomas. *Paediatric Intensive Care, Sheffield Childrens Hospital, Sheffield, UK*

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Paediatric oncology patients form a significant proportion of patients admitted to PICU. We studied the short and long term survival of oncology patients admitted to a tertiary PICU.

Aim To evaluate the short-term and long-term survival of children admitted to PICU with cancer related problems and the impact of clinical variables on survival.

Methods Retrospective analysis of the PICANet data and case records of all oncology patients aged less than 16 years admitted to a tertiary PICU during a 5-year period from 2004 to 2008.

Results A total of 93 patients were identified, 90% survived to discharge from PICU of which 58% survived to 5 years.

64 patients were admitted for post-operative care and 29 for medical reasons. All patients in the post-operative group survived PICU discharge while only 20 patients survived in the medical group; post-discharge mortality at 3 months, 2 years and 5 years was 5, 12, and 3, and 5, 5, and 0 respectively.

Abstract G390(P) Table 1

| Type | Number |
|---------------|--------|
| CNS tumour | 55 |
| Lymphoma | 4 |
| Leukaemia | 12 |
| Solid tumours | 15 |
| Others | 7 |

Twenty patients received mechanical ventilation of which 7 survived; 13 received vasoactive drugs, of which 3 patients survived; 2 received renal replacement therapy but both died.

Nine patients had CNS complications (seizure/raised intracranial pressure), 2 died in PICU and 4 within 3 months of discharge.

Among PICU survivors the duration of stay was one day in 72%; only 3 (5%) received support for more than one organ system.

Among children who died on PICU, 6 died within 24 h; 7 (18%) received support for more than one organ system.

Fifty eight per cent of patients with CNS tumours survived to 5 years. Only 37% of patients admitted with haematological malignancies survived to 5 years.

Conclusions PICU survival for patients admitted solely for post-operative care was better than those with medical complications.

Better survival was associated with fewer days in PICU and fewer organs supported.

The type of malignancy also had a significant impact on both short and long term survival after PICU admission.

G391(P) USE OF NON-INVASIVE RESPIRATORY SUPPORT IN BRONCHIOLITIS: A NATIONAL SURVEY

¹H Turnham, ²R Agbeko, ³J Furness, ⁴A Sutcliffe, ¹P Ramnarayan. ¹*Children Acute Transport Service, Great Ormond Street Hospital, London, UK;* ²*Paediatric Intensive Care, Great North Childrens Hospital, Newcastle-upon-Tyne, UK;* ³*Department of Paediatrics, Darlington Memorial Hospital, Darlington, UK;* ⁴*Institute of Child Health, London, UK*

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Aims Bronchiolitis accounts for a significant proportion of hospital and intensive care admissions in children aged <1 year.^{1,2} Nasal continuous positive airway pressure (nCPAP) has been used traditionally as first-line respiratory support modality.

Heated-humidified High Flow Nasal Cannula (HFNC) therapy is rapidly gaining popularity as an alternate mode of non-invasive respiratory support due to its ease of use. This is despite lack of clinical trial evidence to support its use in bronchiolitis.³ We aimed to establish current practice relating to HFNC and CPAP use in the United Kingdom (UK).

Methods We devised an on-line survey (Survey Monkey, USA) and disseminated the link through regional retrieval services to hospitals with paediatric inpatient facilities. Responses were requested from one senior nurse and doctor from each hospital. The questionnaire covered: hospital characteristics, clinical indications for use of CPAP and/or HFNC in bronchiolitis, and willingness to participate in future research. Denominator data (number of UK hospitals with inpatient services) was obtained from the RCPCH 2013 census.

Results Five regional retrieval services distributed the survey to their local hospitals, covering 109 of 191 hospitals in the UK with inpatient paediatric services. 156 respondents from 102 individual hospitals provided information (response rate: 94%).

Of the 102 hospitals, 23 had a dedicated paediatric intensive care unit, 38 a dedicated paediatric high dependency unit (PHDU), 50 had dedicated PHDU beds on general paediatric wards. 30% have no PHDU provision. 95 (93%) units can deliver nCPAP to children with bronchiolitis and 73 (71%) can deliver HFNC. The main indications for HFNC use: as an alternative to nCPAP (69%), escalation of therapy from low flow oxygen (74%), and weaning from nCPAP (57%). The majority (71%) would choose HFNC over nCPAP in a deteriorating child with bronchiolitis; despite this, 76% were prepared to randomise infants between nCPAP and HFNC in a clinical trial.

Conclusions Use of HFNC is common across UK hospitals. Despite this, survey responses support clinical equipoise amongst the majority of clinicians, indicating the feasibility of a trial of nCPAP and HFNC at this time.

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