

Central Nervous System Tumours in Children Under the Age of Three: A Population Study by the Hong Kong Paediatric Haematology/Oncology Study Group

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Background: Central nervous system (CNS) tumours in children below the age of three represents special challenge to pediatric oncologists. Population-based epidemiological data on this particular patient group is lacking in Hong Kong.

Method: Review of data from a population-based pediatric tumour registry of the Hong Kong Paediatric Haematology/Oncology Study Group between 1999 and 2011.

Results: Eighty-one children with primary CNS tumours from 0-3 years were identified (annual incidence: 4.16 cases per 100,000). Forty-one (50.6%) were male and mean duration of FU was 94 months (± 8.1). Primary tumours were infratentorial in 43 (53.1%). The tumour types in decreasing frequency were astrocytoma (n=17), medulloblastoma (n=16), ependymoma (n=13), CPT (n=7), PNET (n=7), ATRT (n=6), GCT (n=5), craniopharyngioma (n=4) and ganglioglioma (n=3). Three patients presented antenatally. Treatment included surgery in 82.7%, chemotherapy in 50.6% and radiotherapy in 25.9%. There were 29 deaths (35.8%) and 19 relapses (23.5%) during the review period with the 1y-OS, 5y-OS, 1y-EFS and 5y-EFS being 79.4% (± 4.6), 63.5% (± 5.9), 68.9% (± 5.3) and 52.5% (± 5.9) respectively. Significantly better OS and EFS were observed in patients who received gross-total resection but those with high-grade tumours, antenatal diagnosis or ATRT/PNET had worse outcome. Survival did not differ with age. Comparison with statistics from other studies revealed higher rates of embryonal tumour, GCT and craniopharyngioma in Hong Kong Chinese. Disease outcome appeared to be better in our cohort comparing to previous reports probably due to the higher proportion of GCT locally.

Conclusion: We described the epidemiology and treatment outcome of CNS tumours in young children locally; ethnical differences in the frequency of particular tumour types might exist.

Application of High-resolution Anorectal Manometry in Children with Intractable Constipation

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Aim of the study: Chronic constipation can be due to slow colonic transit or sphincter problem or both. The role of sphincter pathophysiology in constipation is still unclear. This study aims to review the application of high-resolution anorectal manometry (HARM) in children with intractable constipation.

Methods: From March 2013 to January 2014, a retrospective review was conducted to study the children aged above 5 years who underwent HARM at our institution. They had either (i) chronic constipation according to the Rome III criteria, or (ii) complications due to fecal retention needing hospitalisation, followed by poor response to a 6-month period of conservative treatment. These patients were compared with the historical control, from September 2011 to February 2013, who received conventional manometry (CARM) by station pullthrough technique.

Main results: Nineteen children (16 boys, median age 9 years) received HARM and 18 children (10 boys, median age 8 years) received CARM.

	HARM	CARM	p-value
Resting anal pressure ≥ 70 mmHg	73.7%	55.6%	0.248
Relaxation of anal sphincter $< 10\%$ on pushing	47.1%	12.5%	0.031

Patients with resting anal pressure ≥ 70 mmHg were considered to have anal sphincter hypertonicity. Patients who had anal pressure relaxation of $< 10\%$ or paradoxical contraction on pushing were regarded to have anal sphincter dysynergia.

In HARM patients with anal sphincter hypertonicity, 66.6% also had sphincter dysynergia, comparing to 0% for those with resting sphincter pressure < 70 mmHg (p=0.012).