A Structured Multi-Disciplinary Team Approach to Pheochromocytoma

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Background

Pheochromocytoma is a rare (0.2/100000) catecholamine-secreting tumour of the adrenal glands leading to haemodynamic instability, especially during intubation, tumour manipulation, and with increased intra-abdominal pressure. Children are more likely to have tumours that are extra-adrenal, bilateral or multiple compared to adults.

Working in a national tertiary level hospital we encounter on average four cases a year (validated audit data). There is currently no guideline for medical and nursing staff as to how to approach the patient pre-operatively or intra-operatively or post-operatively. The aim of this work is to determine if having a disease specific guideline on how to optimise a patient pre-operatively and care for them both during and after surgery improves quality of patient care.

Morbidity and mortality increase by 25 fold for emergency surgery compared to elective.

Method

An extensive literature review focusing on paediatrics was undertaken. A needs-based assessment for the clinical pathway was derived from operating theatre audit data. An evidence-based algorithm was developed in conjunction with intensive care, anaesthesiology, and endocrinology with respect to pre, intra and postoperative care of these patients. Prospective cases were treated according to the algorithm and the intra and post-operative course of patients compared.

Results

Six surgical resections of phaeochromotytomas in 5 patients were undertaken in this institute since 2012. One patient had two surgeries three years apart. Inadequate pre-op blood pressure control accounted for delays in 83.3% of cases. Two patients had inadequate pre-op investigations including no ECG or cardiac echo. An intra-op hypertensive crisis occurred in 33.3% of surgeries. No emergency surgeries were performed. PICU length of stay has remained consistent over time.

Poor BP Control on Pre-Op Anaesthesiology Review

Length of Stay PICU Post-Op

Intra-Op BP Control

Conclusions

The most significant finding was that over 80% of cases were delayed due to poor pre-op deficits. Intra-op BP control can be difficult but all efforts should be made to prevent hypertensive crises. The implementation of an evidence-based clinical pathway for perioperative management of this rare paediatric condition will hopefully improve on these shortcomings.