Harlequin Syndrome Associated with Thoracic Epidural Analgesia

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Two children who presented for surgery received thoracic epidural catheters for postoperative pain management. Postoperatively the patients were found to have unilateral facial flushing with a sharp demarcation along the midline. The rest of their physical examinations were normal. In both patients, the flushing resolved within hours of either decreasing or temporarily stopping the infusion and repositioning the patient or the catheter.

First described by Lance et al in 1988, harlequin syndrome is characterized by a sudden onset of unilateral flushing and sweating of the face and/or upper extremity. Harlequin syndrome results from contralateral blockade of the sympathetic innervation to the vasculature of the face and/or upper extremity. The vasomotor sympathetic fibers of the face and upper extremity leave the cord at T2 to T3 and T4, respectively. The fibers then travel within the sympathetic chain to synapse in the superior cervical ganglion and then within the carotid plexus to reach their effectors. A one-sided sympathetic lesion leads to ipsilateral loss of facial flushing and anhydrosis as a normal response to heat or emotion and excessive sweating and flushing of the contralateral side. Harlequin syndrome is most often idiopathic; however, it can have many etiologies in adults and in children. It is associated with a 64% incidence of abnormal ipsilateral pupils, the majority being miotic, consistent with Horner syndrome. Although harlequin syndrome in this setting is usually transient, it is important to recognize that given the nature of the surgery, harlequin syndrome could be due to surgical nerve injury.

Competing Interests
The authors declare no competing interests.

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References