Harlequin Syndrome after Thoracoscopic Repair of a Child with Tracheoesophageal Fistula (TEF)

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Eur J Pediatr Surg Rep 2019;7:e63–e65.

Abstract

Harlequin syndrome (HS) is a rare dysautonomia of the sympathetic nervous system leading to asymmetric facial flushing and sweating. In the literature, only a few cases of HS after thoracoscopic tracheoesophageal fistula (TEF) repair are reported. We report on a newborn with TEF who developed HS after thoracoscopic repair. On the first day of life, the girl (3,480 g, gestation age: 41 week) underwent thoracoscopic repair of a type C esophageal atresia (TEF; OR time 105 minute) without complications. The postoperative course was uneventful, the patient swallowed and thrived well and did not require esophageal dilations. At 2 years of age, missing facial flushing, transpiration, and warming on the right side of her face during agitation were noticed. As no further intervention was required, the girl and her parents adapted well to the symptoms. Our report shows that the late onset of HS after the surgical procedure is unlikely a direct causal relation to the thoracoscopic operation but rather a shared embryological pathogenesis, like a neurocristopathy.

Keywords

► Harlequin syndrome
► esophageal atresia
► thoracoscopic repair
► neurocristopathy

New Insights and the Importance for the Pediatric Surgeon

Harlequin syndrome is a rare occurrence after esophageal atresia repair with unknown etiology.

Introduction

Harlequin syndrome (HS) is a very rare and seemingly benign condition characterized by unilateral loss of facial sympathetic functions. It was first described by Lance, an Australian professor of neurology, in 1988 in adults who showed unilateral flushing and sweating. He hypothesized that these syndromes occurred due to an idiopathic ipsilateral affection of the sympathetic outflow of the third root.1

Since then, several reports and reviews have been published on this disorder. The cause of HS can be idiopathic and iatrogenic in both adults and children.1–4 Most of the pediatric cases are related to a certain cause, for example, surgical procedure close to the autonomous nervous system in the upper chest, whereas the pathogenesis of congenital cases of the syndrome is still unclear. Leading to disturbance of autonomous functions like sweating and thermoregulation of the skin HS can be regarded as autonomous dysautonomia. It is different from other partial dysautonomias, although etiological and clinical overlap seems possible.5 We report the third case of postoperative HS following thoracoscopic repair of a tracheoesophageal fistula (TEF).6–8

Case Report

A Caucasian baby girl of healthy parents was delivered spontaneously in the 41th week of gestation with a birth weight of...
Figure 1: Girl with a loss of facial flushing and warming of the right side after exertion (age of 3 years; picture shown with parental approval).
nervous system during embryogenesis can result in dysregulated sympathetic innervation. There is evidence for an association of EA and autonomic disturbances such as hyperhidrosis during feeding and hyperthermia. Moreover, the association of EA with certain cardiovascular anomalies may suggest an underlying neurocristopathy of the caudal pharyngeal arch. In this context, one case of HS associated with rare cardiovascular anomalies has been described. It is tempting to speculate that both conditions, EA and HS, could be a result of disturbed neural crest migration. Due to the very limited number of reported cases of HS in children and in particular in association with EA, this conclusion is only hypothetical and further research addressing the pathogenesis of HS is needed.

**Conclusion**

We report the third case of HS in a 3-year-old girl with EA after uncomplicated thoracoscopic repair. The late presentation of the HS in these cases makes a direct causal relation to the surgical procedure implausible. HS may therefore be a (primary) neurocristopathy associated with EA/TEF.

**Conflict of Interest**

None.

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