Harlequin Syndrome in Scoliosis Surgery

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No Disclosures
<table>
<thead>
<tr>
<th>Case 1</th>
<th>Case 2</th>
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<td><strong>M.S., 15y, m.</strong>&lt;br&gt;Congenital scoliosis&lt;br&gt;Hemivertebra T3 left&lt;br&gt;Wedge vertebra C7 and synostosis C4-7&lt;br&gt;Posterior hemivertebra resection T3 including the proximal part of the rib&lt;br&gt;Instrumentation T1 to T5&lt;br&gt;Ligation of the nerve root T3</td>
<td><strong>E.S., 6y, m.</strong>&lt;br&gt;Congenital scoliosis&lt;br&gt;Hemivertebra T3 right&lt;br&gt;Rib synostosis T1/2 left and T4/5 right&lt;br&gt;Posterior hemivertebra resection T3 including the proximal part of the rib&lt;br&gt;Instrumentation T1 to T5&lt;br&gt;No nerve root ligation</td>
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Uneventful postoperative course

Patients were allowed to start with sports activity 3 months postop.
Both patients complained unilateral facial flush when starting with sports (jogging)

Case 1  (HV T3 left)
Flush: right side
Anhydrosis: left side
Pathology: left side

Case 2  (HV T3 right)
Flush: left side
Anhydrosis: right side
Pathology: right side
Harlequin Syndrome

Symptoms:

- Paleness and anhydrosis at the affected side, obvious by a flush at the contralateral side during sports activity

Incidence

- Rare, case reports

Etiology:

- idiopathic
- congenital
- post epidural anesthesia
- post thoracic surgery
Harlequin Syndrome

Anatomy/Pathophysiology:

- Sympathetic trunk: paravertebral ganglion
- Lesion probably at the level T2/T3
Harlequin Syndrome

Literature:

- Harlequin and Scoliosis: none
Harlequin Syndrome

Literature:

- **Idiopathic/ congenital**


Harlequin Syndrome

Literature:

- **Epidural anesthesia**


Harlequin Syndrome

Literature:

- Thoracic surgery


Harlequin Syndrome in Scoliosis Surgery

Discussion:

- Lesion of the sympathetic trunk / paravertebral ganglion ipsilateral to the hemivertebra
  - Approach including costotransversectomy
  - Resection of the vertebral body
  - Resection of the rib head
  - Manipulation within the neuroforamen
  - Ligation of the nerve root (case 1)
  - Stretching of sympathetic fibers
  - Correction maneuver with closing of the gap
Harlequin Syndrome in Scoliosis Surgery

Discussion:

➢ How to avoid …

• Gentle dissection within the foramen
• Avoid ligation of the nerve root
• Dissection of rib head and vertebral body close to the bony surface (leave the periosteum in young children?)
• Especially careful at the level of T2 and T3