

# Harlequin Syndrome in Scoliosis Surgery

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**No Disclosures**



# Harlequin Syndrome in Scoliosis Surgery

## Demographics

### Case1

**M.S., 15y, m.**

**Congenital scoliosis**

**Hemivertebra T3 left**

**Wedge vertebra C7 and synostosis C4-7**

**Posterior hemivertebra resection T3  
including the proximal part of the rib**

**Instrumentation T1 to T5**

**Ligation of the nerve root T3**

### Case 2

**E.S., 6y, m.**

**Congenital scoliosis**

**Hemivertebra T3 right**

**Rib synostosis T1/2 left and T4/5 right**

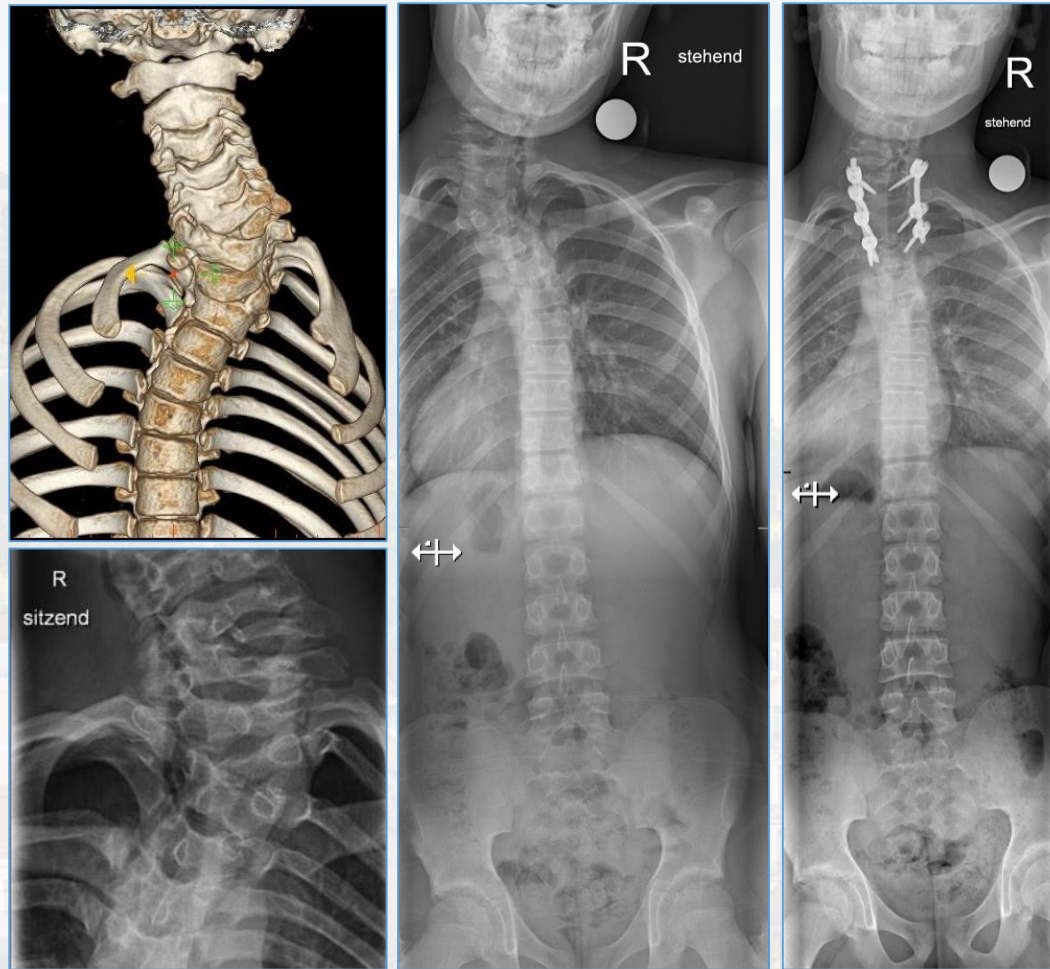
**Posterior hemivertebra resection T3  
including the proximal part of the rib**

**Instrumentation T1 to T5**

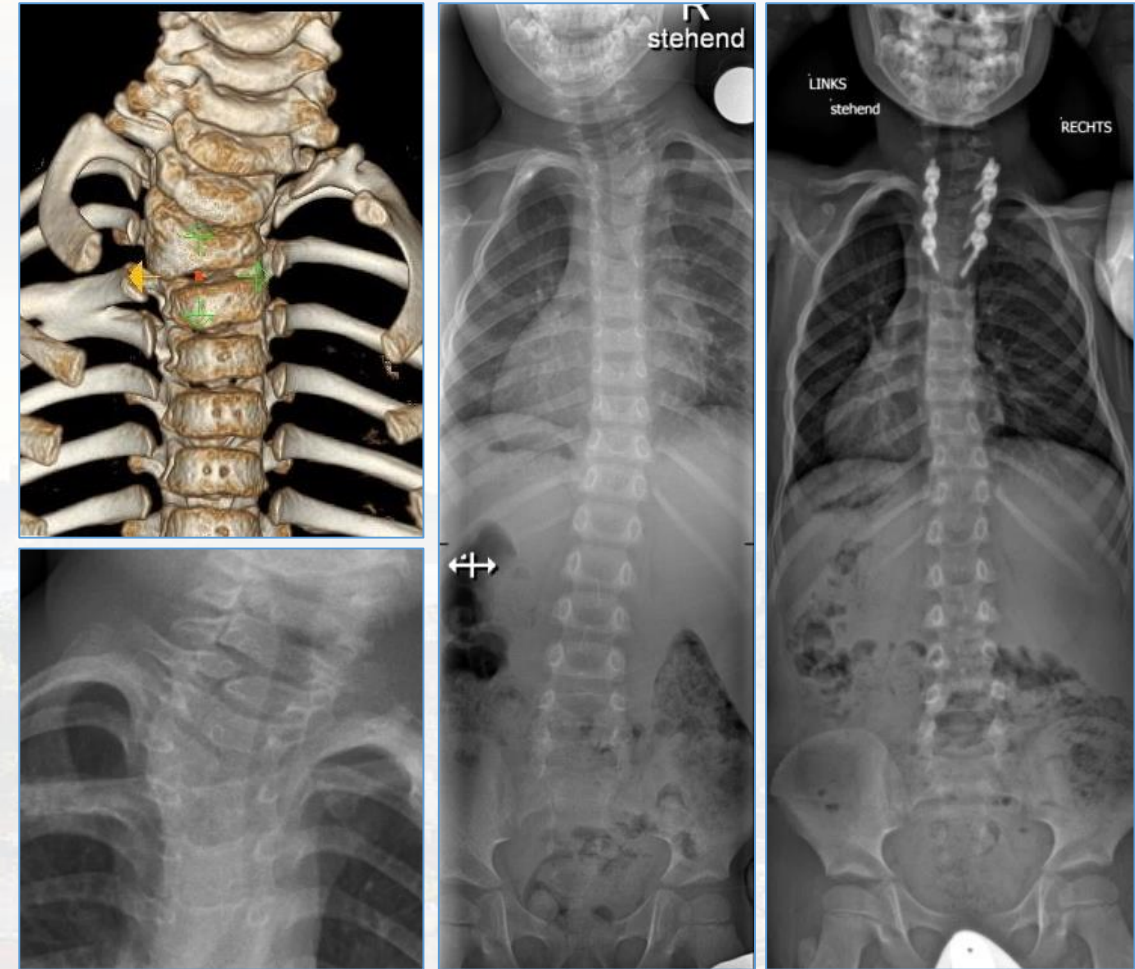
**No nerve root ligation**



## Case 1



## Case 2



**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)**

**Case1 (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:

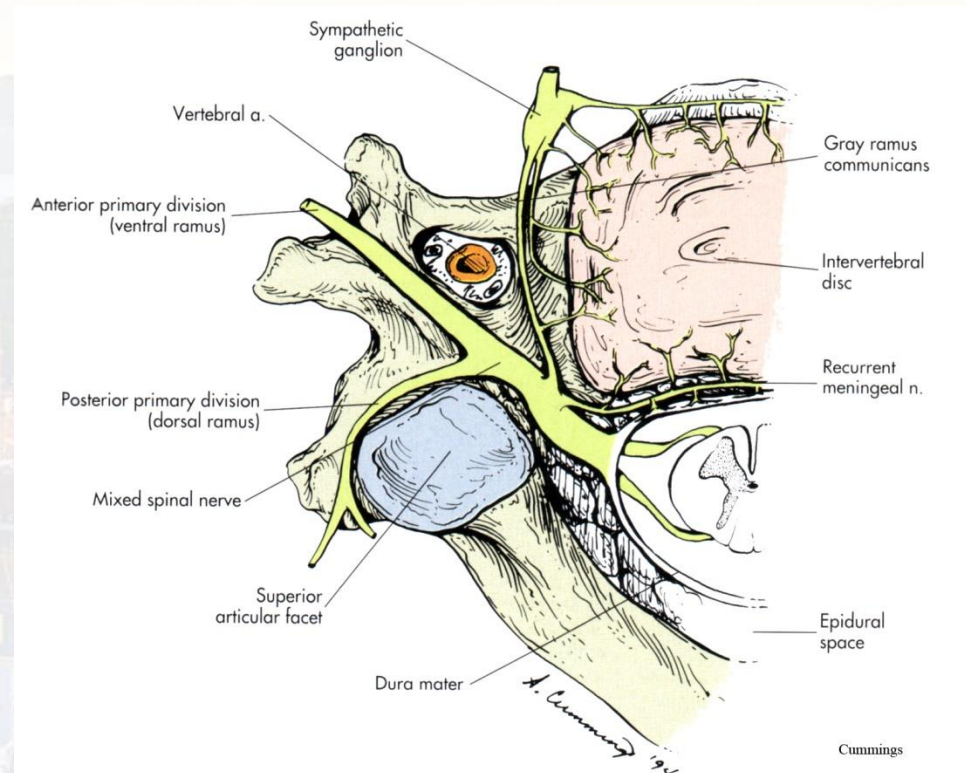
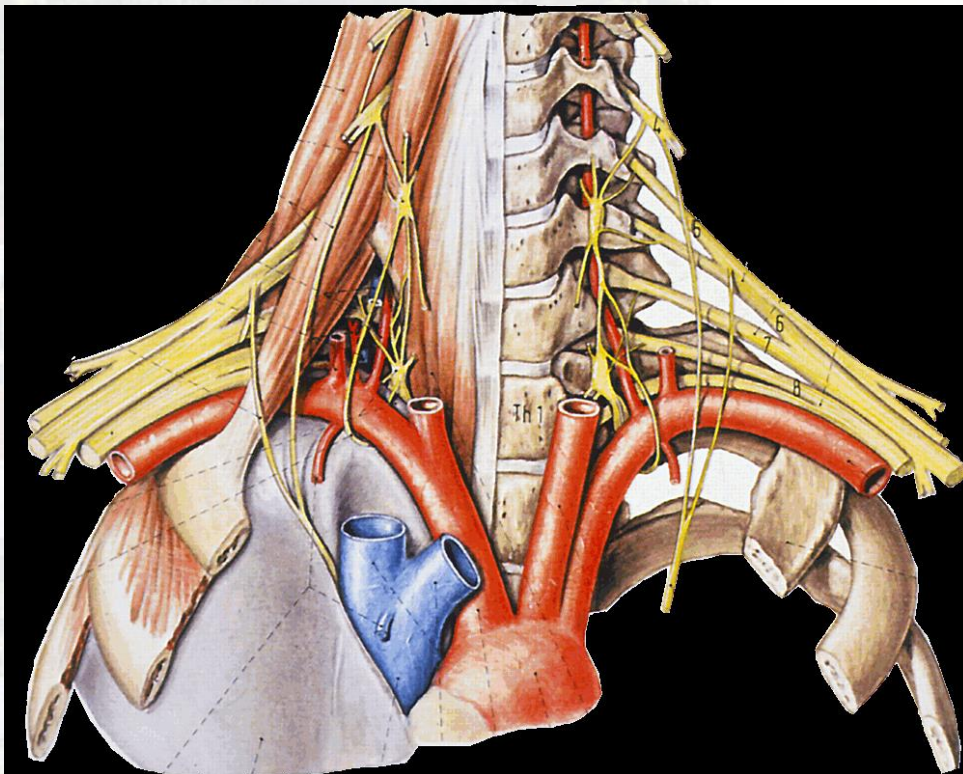
- **idiopathic**
- **congenital**
- **post epidural anesthesia**
- **post thoracic surgery**



# 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

Kang JH, Shahzad Zafar M, Werner KE. Child Neurology: An infant with episodic facial flushing: A rare case and review of congenital harlequin syndrome. Neurology. 2018 Aug 7;91(6):278-281.

Algahtani H, Shirah B, Algahtani R, Alkahtani A. Idiopathic Harlequin Syndrome Manifesting during Exercise: A Case Report and Review of the Literature. Case Rep Med. 2017;2017:5342593.

Kim JY, Lee MS, Kim SY, Kim HJ, Lee SJ, You CW, Kim JS, Kang JH. A pediatric case of idiopathic Harlequin syndrome. Korean J Pediatr. 2016 Nov;59(Suppl 1):S125-S128.

Jain P, Arya R, Jaryal A, Gulati S. Idiopathic harlequin syndrome: a pediatric case. J Child Neurol. 2013 Apr;28(4):527-30.

Lance J W, Drummond P D, Gandevia S C, Morris J L (1988) Harlequin syndrome: the sudden onset of unilateral flushing and sweating. Journal of Neurology, Neurosurgery, and Psychiatry 51:635-642.





# Harlequin Syndrome

## Literature:

### ➤ Epidural anesthesia

Redondo JM, Rivas M, Martín ML, Torrado MD, Rodríguez MJ. Harlequin syndrome as a rare complication after epidural anesthesia in an obstetric patient. *Minerva Anesthesiol.* 2018 Apr;84(4):524-526.

Lefevre A, Schnepfer G. Development of Harlequin Syndrome following placement of thoracic epidural anesthesia in a pediatric patient undergoing Nuss procedure. *Clin Case Rep.* 2017 Aug 3;5(9):1523-1525.

Nagasaka Y, Wasner G, Sharma B, Fleischmann K. Harlequin Syndrome After Thoracic Paravertebral Block. *AA Case Rep.* 2016 Feb 1;6(3):48-51.

Zinboonyahgoon N, Srinivasan S, Narang S. Harlequin Syndrome Following Implantation of Intrathecal Pumps: A Case Series. *Neuromodulation.* 2015 Dec;18(8):772-5.

Boling B, Key C, Wainscott J, Rebel A. Harlequin syndrome as a complication of epidural anesthesia. *Crit Care Nurse.* 2014 Jun;34(3):57-61.



# Harlequin Syndrome

## Literature:

### ➤ Thoracic surgery

Kydes AT, Kelley AM, Pandya SR, Barst SM. Postoperative Harlequin Syndrome: Case Report of a Rare but Clinically Striking Condition. *A A Pract.* 2018 Apr 15;10(8):215-217.

Hussein N, Haqzad Y, Lodhia J, Begum S, Cowen M. Harlequin syndrome following upper lobectomy. *Asian Cardiovasc Thorac Ann.* 2018 Mar;26(3):234-235.

Lee DH, Seong JY, Yoon TM, Lee JK, Lim SC. Harlequin syndrome and Horner syndrome after neck schwannoma excision in a pediatric patient: A case report. *Medicine (Baltimore).* 2017 Nov;96(45):e8548.

Jeon YJ, Son J, Cho JH. Harlequin Syndrome Following Resection of Mediastinal Ganglioneuroma. *Korean J Thorac Cardiovasc Surg.* 2017 Apr;50(2):130-132.

Jung JM, Lee MH, Won CH, Chang SE, Lee MW, Choi JH, Moon KC. Iatrogenic harlequin syndrome: a new case. *Ann Dermatol.* 2015 Feb;27(1):101-2



# 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