

# Stuve-Wiedemann Syndrome (SWS): A Case Report and Series of Anesthetics

Maureen Higgs MD, Tammy Wang MD

## Introduction

- Leukemia Inhibitory Factor Receptor Gene: autonomic nervous system dysfunction with unexplained hyperthermia, disordered breathing, hyperhidrosis, swallowing dysfunction, bowing of the long bones and scoliosis
- Many children die as infants from hyperthermia or disordered breathing
- Anesthesia issues: unexplained hyperthermia seen in most SWS, along with a subset who have inborn errors in the mitochondria of the respiratory chain.

## Case Report

- 4 YO F w/PMHX SWS, severe scoliosis, long limb bowing, and prior OSA transferred from Palestine to LPCH
- PSHX Palestine: Tonsillectomy for OSA, MRI
- Anes Palestine: Used VA without clinical issues
- PSHX LPCH: 2 osteotomies, dental rehab, Mehta Cast
- Anes LPCH: PO midazolam, MH precautions (N2O/PIV/TIVA) CLE for osteotomies, 1 Anes w/VA for induction with planned change to TIVA
- No clinically significant issues during LPCH anesthetics



## Discussion

- MH precautions still in question for these patients given unpredictable hyperthermia, while use of a TIVA may place the subset of SWS patients w/mitochondrial defects at risk.
- Cormier, et al: NL muscle bx in series of SWS patients
- Bonthius, et al: series of sevoflurane anes in a 3 YO, no complications
- LPCH Anes series 4YO w/both VA (Palestine) and TIVA without complications
- More research needed to clarify the safest anesthetic
- Currently, paucity of literature on which to base anesthetics.

## References

- Stuve A, Wiedemann MR. Congenital bowing of the long bones in two sisters. *Lancet* 1971;ii:495.
- Cormier-Daire V, Munnich A et al. Presentation of six cases of Stuve-Wiedemann syndrome. *Pediatric Radiol* 1998;28:776-780.
- Bonthius D, Morava E, et al. Stuve-Wiedemann syndrome and related syndromes: case report and possible anesthetic complications. *Pediatric Anesthesia* 2009; 19: 212-217.