West syndrome (WS) is an X-linked disease associated with epileptic/infantile spasms, abnormal brain wave patterns, and intellectual disability (2). Mutations in the Aristaless related homeobox gene (ARX) are found in patients with this condition. Development of the pancreas, testes, brain, and skeletal muscles may involve the ARX protein (4). In the United States, WS is only seen in 1.5-2.0 live births per 10,000 children aged 10 years or younger. Most deaths related to this disease occur at or before age 10 (3). The therapy for these patients mainly includes medications such as adrenocorticotropin hormone, and conventional antiepileptic drugs (1).

This case report presents anesthesia management in a 12-year old 30 kg female with WS who underwent bilateral ophthalmoscopy and blood work under anesthesia. Patient was a twin gestation born at 34 weeks. Physical exam revealed contraction deformities in all extremities and chest deformity. Her airway appeared grossly normal. She communicated minimally with sign language and was wheel chair bound.

Inhalational mask induction was performed with oxygen, air, and sevoflurane. Laryngeal mask airway (LMA) size #3 without difficulty. Anesthesia was maintained with sevoflurane and 2:1 mixture of air and oxygen. She breathed spontaneously throughout the case. No seizure activity occurred clinically during the preoperative and intraoperative period. Once the bilateral ophthalmoscopy was complete, LMA was removed, and placed her on a venti-mask. No intravenous medications were given during the case. She remained in the recovery room for an extended period for observation. No seizure activity occurred, and she was discharged her home the same day.

These patients are considered possibly difficult intubation due to anatomic malformations. Other issues include difficulty in placing intravenous catheter, positioning challenges, and epileptic seizures management. Tuberous sclerosis is seen in 10-30% of these patients and therefore, thorough preoperative evaluation of cardiac and kidney function is also vital (2). The key in management of these patients is to ensure that the patients are optimized on anti-seizure medications and to avoid conditions that would predispose them to seizures.

References
2. Şahin, Sevtap Hekimoğlu et al. "Anaesthesia Management of a Child with West Syndrome." Turkish Journal of Anaesthesiology and Reanimation 42.6 (2014): 362–364. PMC. And Figure 2.
5. Figure 1: http://www.chp.edu/our-services/brain/neurology/epilepsy/types/syndromes/infantile-spasms