Introduction
Children with Down syndrome (DS), when compared with their age and gender matched counterparts, are more susceptible to OSA (1). Persistent sleep apnea is noted in 30-50% despite adenotonsillectomy (AT) which results in further testing and invasive procedures such as lingual tonsillectomy, craniofacial surgeries and tracheotomy due to poor tolerance to CPAP (2).

We have successfully performed drug induced sleep endoscopy (DISE) and Hypoglossal Nerve stimulator (HNS) placement. This novel procedure to halt the cardiopulmonary and neurobehavioral changes has improved the quality of life in these patients.

Methods
Eligibility criteria for HNS placement include failed/ineffective CPAP or need for tracheotomy, persistent OSA following AT and/or lingual tonsillectomy on a recent polysomnography (PSG).

An anesthesia consult to assess the airway and address perioperative risks related to DS and OSA help minimize postop complications.

DISE is performed using either Propofol or Dexmedetomidine bolus dose followed by maintenance with an infusion which is titrated to mimic natural sleep.

This technique provides optimal conditions for examination of the tongue and observation of dynamic upper airway obstruction. DISE thus helps identify the location of obstruction and quantify the degree of collapse.

Findings on DISE:
Images show retroflexed epiglottis obscuring the vocal cords and base of tongue causing almost complete airway obstruction.

Implantation of the HNS device is performed under GA with intravenous induction; the airway is secured with a nasal endotracheal tube. Long acting neuromuscular blockade is avoided allowing for continuous nerve monitoring during the procedure. Maintenance is achieved with use of a volatile agent or intravenous technique.

Dexmedetomidine administered intravenously during this procedure also offers the added benefits: minimizes the use of opioids, preserves respiratory drive and avoids emergence delirium in patients with OSA.

HNS device is implanted in the chest and the electrodes are sutured to the twelfth cranial nerve (Figure 1).

The external components include physician programmer (A) and patient programmer (B).

The physician programmer communicates with the internal piece through telemetry to adjust device settings. The patient programmer turns therapy on and off and adjusts stimulation amplitude for home use.

Respiratory effort triggers the device, stimulating the hypoglossal nerve to protrude the tongue forward during inhalation and avoid obstruction during sleep (4, 5). This decreases velopharyngeal and oropharyngeal resistance to airflow and results in significant improvements in Apnea Hypopnea Index (AHI) and Oxygen Desaturation Index (OSA) in adults (5).

Preliminary data with HNS placement have shown significant improvements in post procedure PSG in 6 children with DS. One patient with a tracheostomy was successfully de-cannulated once he achieved a low AHI score on PSG.

Conclusion
This new modality may decrease morbidity from long term consequences of OSA and improve the quality of life in adolescents with DS.

References