INTRODUCTION
CHARGE Syndrome (CS), also known as Hall-Hittner syndrome, is a congenital disorder comprised of multiple anomalies (coloboma, heart defect, atresia choanal, retarded growth and development, genital hypoplasia, ear anomalies/deafness). First described in 1979 by Hall and Hittner, it is caused by a mutation in the gene CHD7 on chromosome 8.1 Facial and upper airway features of CS, including midface hypoplasia, micrognathia, cleft lip, and palate, anterior larynx, and subglottic stenosis, make the airway management of these patients a challenge for the anesthesiologist.

Because difficult airway management is a concern, spontaneous ventilation is recommended until the airway is secured. We report the use of dexmedetomidine (DEX) for deep sedation in a CS patient who required nasotracheal intubation using a fiberoptic bronchoscope (FB) while breathing spontaneously.

CASE REPORT
A 13-year-old 25-kg female with CS presented for epiphysiodesis of left femur and tibia. Medical history was significant for reactive airway disease and repair of VSD at 8 years of age. Medications included albuterol, fluticasone, and salmeterol.

The surgery was canceled 12 months prior to this visit because her airway could not be secured by conventional direct laryngoscopy maneuvers performed by two pediatric anesthesiologists. Our approach was to perform a nasotracheal fiberoptic intubation with sedation using DEX.

After placing standard ASA monitors, anesthesia induction was performed using sevoflurane with 50% N₂O in O₂ by mask. After peripheral intravenous access was obtained and inhalation agents were discontinued, DEX 1 μg·kg⁻¹ bolus was given intravenously over 10 minutes followed by a continuous infusion at 0.6 μg·kg⁻¹·hr⁻¹.

At this time, patient remained sedated and ventilating spontaneously. Two cottonoids soaked in 4% cocaine were inserted into the right nostril and removed after 5 minutes. A flexible FB (OD 3.8 mm; Olympus) was inserted into the right nostril and after visualization of the vocal cords, 2 mL of 2% lidocaine was sprayed on the larynx via the suction port. The scope was then passed through the vocal cords until the carina was closely visualized. After a 5.0 mm cuffed ETT was passed over the scope, the FB was removed, the cuff was inflated, and end-tidal CO₂ and bilateral breath sounds were confirmed.

Excellent intubating conditions were achieved, and no hemodynamic changes or patient movement were observed during the tracheal intubation. DEX infusion was then discontinued and anesthesia was maintained with sevoflurane with 50% N₂O in O₂ and vecuronium bromide. At the end of the 3-hour procedure, anesthesia was discontinued and the patient was awakened. When signs of normal motor and ventilatory responses were appreciated, the patient was successfully extubated. She was transported to PACU and met discharge criteria shortly thereafter. On postoperative evaluation, the patient had no anesthesia-related complications.

DISCUSSION
Facial defects present in CS have been known to make securing an airway challenging, with the severity of difficulty increasing as the patient ages.2 Because of the high risk of loss of airway control after anesthesia induction, it is recommended that spontaneous ventilation be preserved during intubation.

Providing optimal conditions for intubation in a child with a known difficult airway is challenging as it requires a deep level of sedation and maintenance of spontaneous ventilation. Current methods of sedation for fiberoptic intubation such as benzodiazepines, propofol, or opioids have their limitations with regard to respiratory depression and loss of airway control.

DEX is a selective alpha-2 agonist that has potent analgesic, anxiolytic, sedative, and antisialagogue properties.3 Spontaneous ventilation is maintained as respiratory depression is minimal. Bradycardia and hypotension, potential side effects of alpha-2 agonist administration, were not observed in our patient. The 4% topical cocaine she was given may have prevented both bradycardia and hypotension. The use of DEX in combination with topical cocaine and lidocaine provided optimal conditions for the airway management in our patient with CS.

REFERENCES