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Anaesthetic management in a case of donohue syndrome for pedodontic surgery - A rare case report

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Abstract

An unusual case of 8 year old female, Donohue syndrome, patient was admitted in Pravara Rural Hospital, Loni, Ahmednagar for multiple teeth extraction under general anaesthesia, presenting unique challenges for peri-operative anaesthetic management.. Patient had depressed nasal bridge with flaring of nostrils was noted. On systemic examination, patient had abdominal distension with everted umbilicus and dilated superficial veins; B/L lower lung fields had decreased air entry with reduced lung volumes. Left nasal intubation with Flexometallic tube attempted but failed. One episode of epistaxis and bradycardia was managed with nasal pressure and inj atropine 0.1 mg iv respectively. Intubation was successful on second attempt with ETT no. 5 passed orally with diligent monitoring for bradycardia. Patient maintained on O_2 + N_2O ratio along with Sevoflurane; inj vecuronium for muscle relaxation. Post-operatively patient had bronchospasm which was managed with adrenaline and duolin nebulisations 4th hourly. Patient was extubated on POD1 after complete resolution of bronchospasm and normal ABG and all routine investigations. Major anaesthetic considerations in Donohue syndrome include difficult airway management, cardiorespiratory abnormalities like cardiomyopathy, cardiomegaly, abdominal distension, and anemia along with perioperative glycemic management. Features like macroglossia, macrognathia, short neck with irregular dentition are predictors of difficult airway and hence equipment for such management should be kept ready at hand. Decreased Total lung capacity and FRC due to abdominal distension with anemia and anticipated difficult intubation require special attention and alertness as

apneic time is reduced. Thorough knowledge of pathophysiology is essential for successful outcome in management of such rare case.

Keywords: Donohue syndrome, Airway management, General anesthesia, Perioperative hyperglycemia, difficult intubation

Introduction

Donohue Syndrome is an Autosomal recessive (AR) genetic disorder with heterozygous insulin receptor gene mutation resulting in hyperinsulinemia with hyperglycemia.[1,2] Children with Donohue syndrome show characteristic features like macroglossia, low set ears, macrognathia with multiple pathophysiologic abnormalities like anemia, thrombocytopenia, cardiomegaly, valvular lesions, cardiomyopathy, abdominal distension.[2,4] Hyperglycemia predisposes to multiple and repeated infections such as multiple caries teeth. A 8 years old female child presented with such complaints requiring multiple teeth extraction under general anaesthesia presenting unique challenges for peri-operative anaesthetic management.

Case Report

An 8 year old female, k/c/o Donohue syndrome was admitted in Pravara Rural Hospital, Loni for dental caries. Patient was diagnosed as Donohue syndrome in 2019 by karyotyping. Patient was LSCS born with 15 days NICU stay for VLBW. Patient had global developmental delay. Diagnosis of Donohue syndrome was made at 7 yrs of age by karyotyping. Patient was on tablet Metformin with pioglitazone OD since 2 years. There was history of frequent upper respiratory tract infection with last episode being 2 weeks prior to surgery. Preoperative vitals were normal with room air saturation of 98%. Patient weighed 10 kg. She had thickened, wrinkled skin with decreased muscles mass, skin creases showed acanthosis nigricans (figure: 1). She had history of dental caries for which she underwent tooth extraction 6 months back which was unsuccessful as the child was not co- opeartive. Multiple cervical lymph nodes were enlarged probably due to dental caries. Airway examination had significant findings of large protuberant tongue with Mallampati score IV, two rows of malaligned teeth. Depressed nasal bridge with flaring of nostrils was noted. On systemic examination, patient had abdominal distension with everted umbilicus and dilated superficial veins; B/L lower lung fields had decreased air entry with reduced lung volumes. Cardiovascular examination was suggestive of cardiomegaly which was confirmed on Chest X ray with reduced lung fields and 2D ECHO revealed LVH. Horseshoe kidney was detected on abdominal USG. Complete blood count revealed Hb of 7.9 g/dl which was optimized to 11 g/dl with preoperative PCV transfusion. Blood glucose level was 503 mg/dl on admission for which H. Actrapid infusion was started. Pre op BSL was 180 mg/dl. Infusion was continued perioperatively.

Written verbal informed high risk consent was taken and patient was kept NBM for 6 hours prior to surgery. Patient was premedicated with inj. glycopyrrolate 0.06 mg iv, inj midazolam 0.4mg iv, inj fentanyl 20 mcg iv and for sedation inj ketamine 5mg iv was given. Monitoring was done with pulse oximetry, NIBP, ECG, and $ETCO_2$. Patient was preoxygenated for 3 min induced with inj propofol 20 mg iv, inj scholine 20 mg iv. Cormack-Lehane Grade was 4 on laryngscopy. Left nasal intubation with Flexometallic tube no. 5.5 was attempted but failed. One episode of epistaxis occurred and was managed with nasal pressure. One episode of bradycardia was managed with inj atropine 0.1 mg iv.

Intubation was successful on second attempt with ETT no. 5 passed orally after repeat doses of propofol and scholine with diligent monitoring for bradycardia. Throat packing was done post intubation to prevent aspiration(Figure 2). Patient was maintained on O_2 + N_2O ratio in 40:60 ratio along with Sevoflurane and MAC was maintained from 1-1.5; inj vecuronium for muscle relaxation. Paracetamol and fentanyl were given for analgesia. RL with 25% dextrose was given for fluid maintainence. Hourly BSL monitoring was done with values being with BSL value ranging between 180-200 mg/dl. Infusion of H. Actrapid 0.12U/kg/hr was continued. Surgery lasted for three and half hours and multiple teeth were extracted. As there were multiple attempts at intubation with associated nasal bleed and extensive nature of procedure, elective ventilation was considered and patient was shifted to PICU intubated. Dexamethasone and Hydrocort were given iv to limit post op oral and nasal edema.

Post-operatively patient had severe bronchospasm in right middle and lower lobes which was managed with adrenaline and duolin nebulisations 4^{th} hourly. Patient was extubated on POD1 after complete resolution of bronchospasm and normal ABG and all routine investigations.

Discussion

In this case due to dysmorphic facial feature associated with macroglossia and large jaw there was difficult intubation scenario which was anticipated and difficult airway cart was kept ready. Presence of cardiomegaly with LVH increases risk of adverse cardiac events which might explain bradycardia after first attempt at intubation. Thrombocytopenia with coagulation abnormalities is also associated with Donohue syndrome and presence of distorted facial anatomy might explain epistaxis, though the coagulation profile was normal in our patient.Post operative incidence of bronchospasm could be due to seepage of blood and debris in the tracheal lumen after removal of throat pack as uncuffed tube was used. Also the history of URTI increases risk of peri-operative bronchospasm and laryngospasm. All these challenges were encountered and managed successfully.

Major anaesthetic considerations in Donohue syndrome include difficult airway management, cardiorespiratory abnormalities like cardiomyopathy, cardiomegaly, abdominal distension, and anemia along with perioperative glycemic management. Features like macroglossia, macrognathia, short neck with irregular dentition are predictors of difficult airway and hence equipment for such management should be kept ready at hand. Decreased Total lung capacity and FRC due to abdominal distension along with anemia and anticipated difficult intubation require special attention and alertness as apneic time would be reduced. Perioperative insulin infusion with monitoring of serum electrolytes is an essential part of anaesthethic management. Cardiomegaly predisposes to ischaemia if myocardial workload increases and hence tachycardia, hypertension, acidosis and hypoxia should be avoided. Thorough knowledge of pathophysiology is essential for successful outcome in management of such rare case.

Conclusion

Donohue syndrome is a rare disorder with altered metabolic, developmental, hemodynamic, cardiovascular and respiratory physiology with important anesthetic considerations like hyperglycemia, anemia, difficult intubation, reduced cardiovascular and respiratory reserve requiring special anesthetic management to ensure optimal outcome.

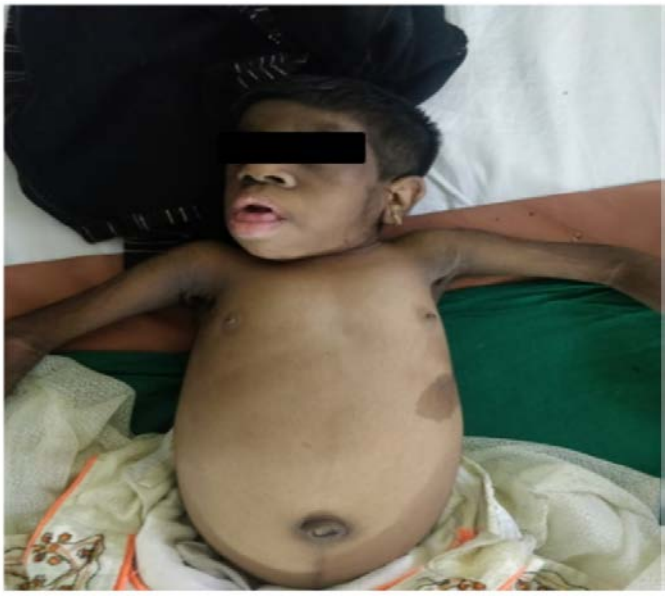

Figure 1

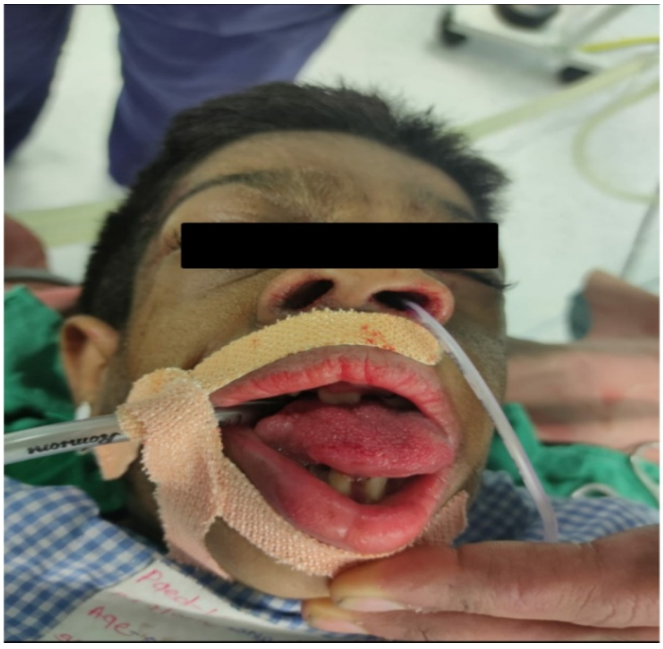

Figure 2

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