Case Report

# A rare case report of anaesthetic management in a paediatric patient with Christ-Siemens-Touraine syndrome

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## Abstract

Ectodermal dysplasia (ED) is a heterogeneous group of disorders which may involve teeth, skin and appendageal structures including hairs, nails, eccrine and sebaceous glands. As ED is very rare in occurrence, the available literature is scarce. We are describing a case of 3 year old male child with suspected foreign body (nose) posted for emergency nasal endoscopy. Anticipating difficult airway, frequent pulmonary infections, temperature monitoring are important considerations in anaesthesia management in a case of patient with Ectodermal Dysplasia. Thorough knowledge about Ectodermal Dysplasia is important to an Anaesthesiologist to tackle related complications in emergency scenarios.

**Key Words:** Christ-Siemens-Touraine syndrome, Ectodermal Dysplasia, emergency surgery, paediatric age group.

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## INTRODUCTION

Ectodermal Dysplasia (ED) is rare group of genetic disorders which occurs due to defect in tissue of Ectodermal origin. It may involve defects in 2 or more of the following: teeth, skin and appendageal structures including hairs, nails, eccrine glands and sebaceous glands. Around 170 variants have been described in literature. [1] Most common variant is anhidrotic E D which is X-linked recessive and characterised by triad of anhidrosis/hypohidrosis, anodontia (i.e. peg shaped teeth) and hypotrichosis. This case report involves anaesthesia management of a 3 years old child with anhidrotic E D with suspected rhinolith or foreign body in nose posted for diagnostic nasal endoscopy under general anaesthesia. The main anaesthetic considerations in these cases are hyperthermia, pulmonary infections and possibility of difficult airway due to deformed teeth.

## CASE REPORT

A 3 year male child weighing 10 Kg presented with history as given by mother suggestive of foreign body in nose with fowl smelling discharge since 10 days. Patient also had history of cough since 1 day. Patient was a diagnosed case of anhidrotic Ectodermal Dysplasia. There was no significant past surgical history. On examination patient had sparse hairs on scalp, periorbital hyper pigmentation, pegged teeth, flattened nasal bridge and dry skin. Fowl smelling discharge through left nostril was present. On external examination no foreign body was visible. Patient was anaemic with haemoglobin 9.6 gm/dL. Other investigations were within normal limit. Decision was taken to give general anaesthesia with endotracheal intubation and controlled ventilation on emergency basis. Difficult airway cart was kept ready in operation theatre with good suction. Patient was nebulised with bronchodilator preparation. Child was playful and cooperative. Premedicated with injection Glycopyrrolate 40 mcg and injection Hydrocortisone

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20mg was given. Patient was transported into operation theatre and routine monitoring devices i.e. Spo2 probe, ECG and NBP were attached. Patient was preoxygenated with 100% oxygen for 3 minutes. Patient sedated with injection Midazolam 0.2 mg intravenously. Patient was induced on inhalational anaesthetic agent sevoflurane. Endotracheal intubation was facilitated with injection Atracurium 5mg. Patient was intubated with uncuffed endotracheal tube no. 4.5 and endotracheal placement was confirmed with end tidal Co2 monitoring. Patient was ventilated with Jackson- Rees circuit. Patient's body temperature was monitored with rectal placement of temperature probe. Temperature was found to be 36.8$^{o}$C. Body temperature was monitored continuously and maintained in the range of 36.1 to 37.0$^{o}$C. Rhinolith was found in the left nostril on nasal endoscopy which was removed uneventfully. This patient possibly developed rhinolith due to thick and inspissated secretions. Endotracheal suctioning was done intermittently during the procedure. The patient was reversed with injection Neostigmine 0.5mg and injection Glycopyrrolate 80 mcg. Patient was extubated after return of normal muscle power, tone and reflexes.

Figure 1

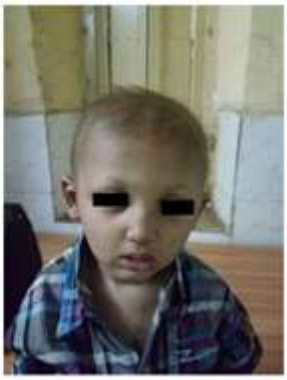

Figure 2

**Legend**

**Figure 1:** Picture showing anodontia (i.e. 'peg shaped' teeth)

**Figure 2:** External appearance of patient with Ectodermal Dysplasia showing hypotrichosis(Sparse hairs on scalp and eyebrows, periorbital hyper pigmentation, depressed nasal bridge)

## DISCUSSION

Ectodermal Dysplasia are rare group of disorders which involve abnormality of ectodermal structures with its reported incidence being 1:100000[1]. Affected individual presents with defects involving teeth, skin and its appendages (hairs, nails, sweat glands etc). 170 such heterogeneous disorders have been described and 30 have been genetically defined.[2] Anhidrotic Ectodermal dysplasia is characterised by triad of features including partial or complete absence of sweat glands, abnormal dentition and hypotrichosis. Genetically it may transmit as X-linked recessive, autosomal dominant or autosomal recessive. There is possibility of difficulty in airway access due to defective dentition. The difficulty is further accentuated as this is paediatric airway. We prepared difficult airway trolley taking into consideration such possibility of difficult airway. Another problem encountered in such cases is hyperthermia due to absence of sweat glands. We strictly monitored body temperature during the procedure to avoid hyperthermia. Respiratory infections and reactive airway are also major concerns while managing such cases. These patients are more prone for bronchospasm and/or laryngospasm due to frequent upper and lower respiratory tract infections. In our management we used nebulisation with bronchodilator preoperatively. Also intravenous steroid was administered to minimize risk of respiratory complications. Very few case reports were found in literature describing management of E.D. Elif *et al* in their case report described successful anaesthetic management of 10 year old child posted for emergency appendicectomy using combination of general and epidural anaesthesia.[3] In their management they highlighted importance of preparation for difficult airway cart. Elif *et al* recommended use of regional anaesthesia in such cases and also emphasized need for continuous temperature monitoring. Sugi *et al* in their case study described epidural anaesthetic in a 20 year old male patient with Hypohidrotic E.D. posted for skin grafting and debridement.[4] Hotta *et al* described successful anaesthesia management of a 10 year old girl child posted for ophthalmic surgery. They emphasized key factors for successful anaesthesia management are airway hydration, monitoring body temperature and preparation for difficult airway cart.[5] Many of the reported cases till today were elective cases. Ours as suspected foreign body in upper

airway was an emergency. Also there are very few cases which describe anaesthesia management in paediatric patient with E D. Airway preparations should be adequate with preoperative nebulisation to humidify airway. It is important to have difficult airway cart ready as intubation of trachea might be difficult in these patients. Temperature monitoring during procedure is highly recommended for early detection and prevention of hyperthermia. Paediatric patients are more prone for development of temperature abnormalities. Vigilant monitoring is needed in paediatric patients.

## CONCLUSION

Knowledge of associated problems in patients with ED is important to avoid complications. Airway humidification, preparing difficult airway cart and temperature monitoring are mainstay in anaesthetic management of patients with ED.

## REFERENCES

1. Palit AIA. What syndrome is this? Pediatr Dermatol. 2006;23:396-8
2. Joseph G Morelli. "Ectodermal Dysplasia" In: Kliegman, Robert, and Waldo E. Nelson: Nelson Textbook of Paediatrics. Philadelphia: Saunders, 2007;18:2666-2667
3. Elif Oral Ahiskalioglu, Ali Ahiskalioglu, Binali Firinci, Aysenur dostbil, Mehmet Aksoy: Anesthetic management of a pediatric patient with Hypohidrotic Ectodermal dysplasia undergoing emergency surgery: Rev Bras Anestesiol. 2015;65(6):522-524
4. Sugi YHK, Suzuki Y, Shono S, et al. Anesthetic management of a patient with Hypohidrotic Ectodermal dysplasia. MASURI. 1999;48:414-6
5. Hotta MKT, Umemura N, Takino Y, et al. Anesthetic management of a patient with Hypohidrotic Ectodermal dysplasia. Masuri. 2000; 49:414-6.

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