

## Brain Tumor in a Case of Down's Syndrome: Anaesthesia Perspectives

Santanu Bora; DNB, Mukul Jain; MD, Arvind Arya; MD and Deepak K Jha; MCh\*

Departments of Neuroanaesthesia and Neurosurgery, Institute of Human Behavior and Allied Sciences, Delhi, India

\*Corresponding author: Deepak Kumar Jha, Departments of Neuroanaesthesia and Neurosurgery, Institute of Human Behavior and Allied Sciences, Delhi, India, Tel: +919868527900; E-mail: [jhadeepak2@rediffmail.com](mailto:jhadeepak2@rediffmail.com)

Received date: May 11, 2016; Accepted date: July 02, 2016; Published date: July 07, 2016

Copyright: © 2016 Bora S, et al. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

### Abstract

Down's syndrome patients are known to have various associated airway and craniovertebral junction anomalies which pose a challenge for anaesthetic management of these patients undergoing brain tumour surgery. Authors report anaesthetic management of a patient of Down's syndrome undergoing brain tumor surgery who had airway of Mallampati class IV with large tongue, small mouth opening and high arched palate with review of relevant literature.

**Keywords:** Down's syndrome; Brain tumor; Difficult airway

### Introduction

Down's syndrome is the commonest congenital syndrome occurring 1 in 800 live births [1]. Atlantoaxial instability (AAD), facial dysmorphism, tracheal stenosis, hypotonia and associated congenital anomalies of airway like cleft lip/palate makes these patients' airway difficult from anaesthesia point of view. Associated cardiac anomalies, pulmonary hypoplasia, depressed immunity status makes their anaesthetic management more challenging. A higher risk of leukaemia is well known in Down's syndrome but brain tumours are rarely reported [2]. Anaesthetic management of a case of Glioblastoma Multiforme (GBM) in a patient of Down's syndrome with difficult airway is reported with review of literature.

### Case Report

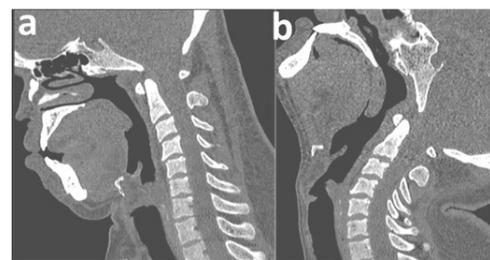
Twenty-nine year old male patient, a known case of Down's syndrome (Based on genetic counselling) was scheduled for craniotomy and excision of right parietal space occupying lesion (SOL). Patient had history of headache and vomiting since 15 days with minimal weakness of left upper and lower limbs. Clinical examination revealed motor power grade IV in both upper and lower limbs on the left side. He had one episode of unconsciousness 10 days back for 2-3 minutes. His other siblings are normal. No other family members had history of brain tumor. He was cooperative but sometimes found difficulty in following instructions and refused to stay without his kin. His intelligence quotient (IQ) was 50-55. He had the history of snoring during sleep.

On examination patient weighed 62 kg with typical features of Down's syndrome. Airway examination revealed macroglossia (Figure 1), high arched palate, Mallampati class IV, mouth opening 3 cm, short neck and restricted flexion of neck. Examination of Cardiovascular and respiratory system revealed no abnormal finding. Serological markers for HIV, HBsAg and HCV were negative. Contrast enhanced magnetic resonance imaging (MRI) study revealed solid cystic intra-axial mass lesion in right parietal lobe with perilesional oedema with foci of calcification, haemorrhage with enhancement characteristics. Dynamic Computed Tomography (CT) scan of cranio-vertebral junction (CVJ) (in flexion and extension) revealed no AAD (Figure 2).

Two-D Echocardiography and electrocardiography (ECG) findings were normal.



**Figure 1:** Shows photograph of patient with macroglossia.



**Figure 2:** Computed tomography sagittal images with bone window settings (a) in flexion and (b) in extension, showing no atlanto-axial instability.

On the morning of surgery, patient's brother was kept with him in the pre-operative area to allay his apprehension and intravenous (IV) cannulation was done. Patient was pre-medicated with IV Glycopyrrolate 0.2 mg, Midazolam 1 mg and Fentanyl 2 mcg/kg. Patient was shifted to operation room (OR) after the patient was sedated. ECG, non-invasive blood pressure (NIBP) and oxygen saturation (SPO<sub>2</sub>) monitorings were done in the OR. Patient was induced with IV Propofol 2 mg/kg and intubated with cuffed flexo-metallic endotracheal tube (7 mm internal diameter) with the aid of

flexible fibreoptic bronchoscope. Muscle relaxation was achieved with IV Vecuronium 0.1 mg/kg. Bronchoscopy revealed large epiglottis (Figure 3) with normal trachea. Anaesthesia was maintained with oxygen (O<sub>2</sub>) and nitrous oxide (N<sub>2</sub>O) and Sevoflurane 0.8%. Invasive blood pressure monitoring was done by cannulating right radial artery and central vein was accessed through right subclavian vein. Patient was positioned supine with head end elevated to 30° and head turned 15° towards left side. Intra-operatively, patient remained hemodynamically stable. Right parietal craniotomy and along with intra-tumoral decompression of the tumor was done. Frozen section examination was consistent with high grade astrocytoma. Patient was extubated at the end of surgery after reversal of muscle relaxation and was kept in ICU for observation for 24 hrs, which was uneventful. Histopathological examination was consistent with Glioblastoma multiforme (GBM), WHO grade IV.



**Figure 3:** Bronchoscopic image of larynx showing enlarged epiglottis (arrow) and normal vocal cords (arrow head).

## Discussion

Craniofacial abnormality is a common finding in Down's syndrome which presents as difficult airway when these patients present for surgery. Factors affecting upper airway include hypotonia, obesity, mid facial hypoplasia, glossoptosis, increased secretions and large adenoid and tonsils [3]. All these factors lead to difficult mask ventilation, difficult laryngoscopy and intubation. They are prone to develop obstructive sleep apnoea and sleep induced ventilator dysfunction, which is exaggerated by narcotic administration and residual anaesthetic concentration in body. They also have increased chances of developing pulmonary vascular disease [4]. Our patient had large tongue, high arched palate and small mouth opening. No sleep apnoea was noted during the observation in ICU for 24 hours after surgery.

Upper airway is a source of obstruction as there is subglottic stenosis, [5,6] tracheal stenosis [7,8] and tracheal bronchus [9] in children with Down's syndrome. These patients should be intubated with 0.5-1.0 mm diameter smaller tube than standard age appropriate endotracheal tube size. Congenital "hour-glass" tracheal stenosis has been described in four patients by Wells and co-workers [8]. Fibreoptic view showed large epiglottis but there was no tracheal stenosis in our case.

Two percent children with Down's syndrome have AAD and cord compression, creating symptoms, whereas 20% have AAD without any symptoms [9,10]. Instability may be due to excessive laxity of posterior

transverse ligament of atlas and malformation of odontoid bone. Atlanto-dental interval of 3-5 mm is considered to be borderline, whereas 12-13 mm is usually associated with symptoms [11]. Preoperative evaluation of CVJ should be done and utmost care should be taken while positioning these patients while surgery and intubation [12]. Our patient had no evidence of AAD in CT scan of CVJ.

Down's syndrome patients have increased incidence of pulmonary infections and relatively increased frequency of positive hepatitis associated antigen [13]. This can be attributed to thymus dependent immune system depression in these patients [14]. Strict aseptic precaution and evaluation of serological markers for viral diseases like HIV, HBsAg and HCV should be done in these patients, which were negative in our patient. Endocardial Cushion defects have been reported in these patients, which were absent in our patient [7].

Patients with Down's syndrome display a unique spectrum of malignancies; with a 10-20 fold increased risk of acute leukaemias and decreased incidence of solid tumours [2]. The basis of decreased solid tumors in these patients is unknown. Satge et al reported 38 cases of central nervous system tumors in Down's syndrome patients and as per their report; GBM is a rare tumor in these patients [2].

We feel that due to the presence of number of factors which pose anaesthetic challenges in these patients, we should approach these patients with caution and use advanced techniques like video-laryngoscope or flexible fibreoptic bronchoscope assisted intubation.

We conclude that Down's syndrome patients are unique in their presentation and impose specific anaesthetic considerations in terms of difficult airway, associated congenital anomalies, congenital heart diseases and depressed immune status. They need special care and emotional support due to their impaired comprehension. Their complex physiology and airway anatomy makes their anaesthetic management challenging and little extra vigilance can avoid complications.

## References

1. Yang Q, Rasmussen SA, Friedman JM (2002) Mortality associated with Down's syndrome in USA from 1983-1997: a population based study. *Lancet* 359: 1019-1025.
2. Satgé D, Monteil P, Sasco AJ, Vital A, Ohgaki H, et al. (2001) Aspects of intracranial and spinal tumors in patients with Down syndrome and report of a rapidly progressing Grade 2 astrocytoma. *Cancer* 91: 1458-1466.
3. Doull I (2016) Respiratory disorders in downs syndrome: Overview with diagnostic and treatment options. *Downs Syndrome Medical Interest*.
4. Chi T, Krovetz LJ (1975) The pulmonary vascular bed in children with Down syndrome. *J Pediatr* 86: 533-538.
5. Miller R, Gray SD, Cotton RT, Myer CM, Neterville J (1990) Subglottic stenosis and down syndrome. *Amer J Otolaryngol* 11: 274-277.
6. Myer CM (1991) Smaller average caliber of the subglottic region in children with Down's syndrome. *Clin Pediatr (Phila)* 30: 646.
7. Shapiro NL, Huang RY, Sangwan S, Willner A, Laks H (2000) Tracheal stenosis and congenital heart disease in patients with Down syndrome: diagnostic approach and surgical options. *Int'l J Pediatr Otorhinolaryngol* 54: 137-142.
8. Wells TR, Landing BH, Shamszadeh M, Thompson JW, BoreKE, et al. (1992) Association of Down syndrome and segmental tracheal stenosis with ring tracheal cartilages: a review of 9 cases. *Pediatr Pathol* 12: 673-682.
9. McLaughlin FJ, Strieder DJ, Harris GB, Vawter GP, Evaklis AJ (1985) Tracheal bronchus: association with respiratory morbidity in childhood. *J Pediatr* 106: 751-755.

- 
10. William C (1998) Atlantoaxial Instability: What Next? *Arch Pediatr Adolesc Med* 152: 119-122.
  11. Alvarez N, Kao A, Schneck MJ (2016) Atlantoaxial Instability in individuals with Down syndrome.
  12. Pushchel SM, Findley TW, Furia J, Gallagher PL, Scola FH, et al. (1987) Atlantoaxial instability in Down's syndrome: roentgenographic, neurological and somatosensory evoked potential study. *J Pediatr* 110: 515-552.
  13. Sutnick AI, London WT, Blumberg BS, Gerstley BJS (1972) Anicteric hepatitis with Australia antigen in patients with Down's syndrome. *Amer J Clin Pathol* 57: 2-12.
  14. Levin S, Schlesinger M, Handzel Z, Hahn T, Altman Y, et al. (1979) Thymic deficiency in Down's syndrome. *Pediatrics* 63: 80-87.