

Anaesthetic Management of a Case of Down Syndrome with Chronic Kidney Disease Undergoing Major Orthopaedic Surgery

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ABSTRACT

Background: Down syndrome is one of the common congenital chromosomal disorders. Renal complications in Down syndrome patients are rare but is increasing in number due to increased survival. Anaesthesia in such patients is challenging due to intellectual disability, anatomical complications and multiple comorbidities.

Case: This case report describes the pre-operative evaluation and anaesthetic management of a 19-year-old male patient with Down syndrome with hypothyroidism with Chronic Kidney Disease stage 4 who underwent open reduction internal fixation with plating for bilateral subtrochanteric fracture under general anaesthesia. It was a complicated surgery for both surgeon and anaesthetist because the patient was anaemic with significant anticipated blood loss along with fluid restriction during the perioperative period. We had major challenges like patient positioning, significant blood loss, hypotension, and early fluid overload. But an extensive preoperative evaluation, optimization prior and proper coordination we overcame the challenges.

Conclusion: Major orthopaedic surgeries in patients like Down syndrome with chronic kidney disease create a major anaesthetic challenge due to expected significant blood loss, chances of fluid overload, multiple comorbidities and intellectual disability. Thorough preoperative evaluation, intraoperative monitoring as well as postoperative period is important as several complications are envisaged. So a closed group management with orthopaedic surgeon, nephrologist, anaesthesiologist, endocrinologist, nursing team is required to handle such complicated cases.

Keywords: Downs syndrome; chronic kidney disease; subtrochanteric fracture; open reduction and internal fixation;

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INTRODUCTION

Down syndrome is by far the most common and best-known chromosomal disorder in humans and the most common cause of intellectual disability. It is primarily caused by trisomy of chromosome 21 due to abnormal cell division, which gives rise to multiple systemic complications as part of the syndrome. It occurs in about 1 in 1,000 babies born each year.¹ Down syndrome nearly always have poor developmental milestones, intellectual disabilities (IQ of age of 8 or 9 years on average) with multiple complications like congenital heart defect, epilepsy, leukaemia, thyroid diseases, gastrointestinal issues and mental disorders.² Although renal disease has been thought to be quite rare the incidence of renal and urological anomalies has been estimated in the range of 3.5–21.4%.³ Pre-existing chronic kidney disease increases perioperative risk.⁴

During general anaesthesia, the most common causes of morbidity are airway and respiratory complications. In Down's

syndrome there are many abnormalities which affect the face and skull: those affecting the airway include a flattened nasal bridge, high arched palate, short nose, small mouth, short neck and spontaneous atlanto axial dislocation. Tracheal intubation can be difficult.⁵ The aim of the case report is to present that Down syndrome which itself has many difficulties for anaesthesia due to anatomical changes and along with hypothyroidism and Chronic Kidney Disease undergoing major orthopaedic surgery involving significant blood loss can be managed successfully without any perioperative complication if you optimize the patient perioperative in coordination with surgeon.

CASE

Informed written consent taken from next of kin for publishing the case report. A 19 year old male patient who weighed 30 kg, a known case of down syndrome with history of ventricular septal defect status post operated with seizure

disorder with Chronic kidney disease stage 4 (non-oliguric and non-dialysis dependent). The patient was accepted in American Society of Anaesthesiologists (ASA) physical status III and was posted for bilateral Open reduction internal fixation with plating for bilateral subtrochanteric fracture.

He was thinly built, poorly nourished weighed only 30 kg. He is the second child to the parent from a non-consanguineous marriage. He was diagnosed as a case of Down syndrome with hypothyroidism with chronic kidney disease (for past 8 years) with seizure disorder and was under medications. He sustained injury by fall following an episode of seizure and had a bilateral subtrochanteric fracture. He was planned for open reduction internal fixation with plating bilateral. On examination the patient had syndromic facies with low set ear, depressed nasal bridge, high arched palate with mallampati class III which suggested possibility of difficult bag mask ventilation, laryngoscopy, and intubation. In addition, the difficult airway typically becomes progressively worse with increasing age (Figure 1). He had history of ventricular septal defect closure in 2014. On auscultation a systolic murmur of grade 2 was noted whereas other areas of chest were clear. He was anaemic with baseline creatinine 3.9 mg/dl. Nephrology consultation was taken for chronic kidney disease and advised to avoid nephrotoxic drug and haemodialysis if required in postoperative period.

The patient was ill looking with bed bound status without any history of fever or upper respiratory tract infections. On ultrasound examination he had bilateral grade 4 hydronephrosis with dilatation of bilateral ureters. As such patients can have anaemia and platelet abnormality, complete blood count showed haemoglobin 10.1 gm%, plt 2.1 lakh/cu mm, Prothrombin time Internationalised normal ratio (PT INR) 1.57, urea 113mg/dl, sodium 142 mEq, potassium 5.3mEq, thyroid stimulating hormone 6.44 U. A written informed parental consent was taken after discussing risks and management of hypotension, blood loss and fluid overload with need of dialysis postoperatively. In view of uncooperative patient due to poor mental status, general anaesthesia was planned.

start of procedure. Patient was given injection tranexamic acid 450 mg intravenous over 15 min to minimize the bleeding. Patient was maintained with restricted fluid therapy to avoid fluid overload.



Figure 1. a) High arched palate, b) Low set ears, flattened nasal bridge

Blood loss was about 500 ml and which was managed with 500 ml crystalloid and 1 unit of packed RBC (280 ml) to maintain the stability of hemodynamics without fluid overload. Anaesthesia was maintained with air, oxygen and sevoflurane with MAC between 0.6 – 0.8 with atracurium in maintenance doses (Table 1). The patient's procedure and anaesthetic courses were uneventful. The patient was reversed and extubated and shifted to the recovery room fully awake state after surgery. Postoperative serum creatinine was raised to 3.86mg/dl and nephrology consultation was taken who advised for conservative management. Patient was discharged after 10 days of hospital admission.

DISCUSSION

The incidence of renal and urological involvement in downs syndrome is high enough to encourage systematic screening. A variety of renal involvement in downs syndrome have been described, including ureteropelvic junction obstruction, vesicoureteral reflux, renal hypoplasia, obstructive uropathy and posterior urethral valves hypospadias, asymptomatic renal pelvic dilatation and asymptomatic kidney ectopia.⁶ Thus, ultrasound should be performed in these infants

Table 1. Haemodynamic and anaesthetic parameters during intraoperative monitoring

Parameters	Baseline	15 min	30 min	45 min	60 min	90 min	120 min	150 min	180 min	210 min	240 min
HR	106	105	105	100	104	98	100	105	108	110	106
BP	110/74	98/60	110/70	116/84	120/76	122/76	120/75	118/76	120/80	120/84	125/86
SPO ₂ (%)	98	100	100	100	100	100	100	100	100	100	100
RR	16	16	16	16	16	16	16	16	16	16	16
MAC	-	0.7	0.6	0.7	0.7	0.8	0.8	0.6	0.6	0.6	0.6

HR: hearth rate; BP; Blood pressure; RR: Respiration rate; MAC: minimum alveolar concentration

After all preparations patient was wheeled into operation room and standard ASA monitoring was established. Besides this entropy electrodes were also attached to maintain accurate minimum alveolar concentration (MAC). After premedication and induction, atracurium was used for intubation and patient was intubated with 6.5 mm internal diameter oral endotracheal tube. Blood product was made available prior to

early in life and, if abnormal, a MCU to rule out any urological abnormality. Approximately one-half of individuals with downs syndrome have congenital heart disease. Endocrine abnormalities in downs syndrome include thyroid dysfunction and type 1 diabetes. Autoimmune thyroid disease is uncommon in young patient with downs syndrome but is common after 8 years of age. Consequently, annual screening is important.

Down syndrome has multiple complications involving different systems such as cardiovascular, respiratory, neurology, haematology, endocrine, gastrointestinal and renal system.⁷ Renal disorders in down syndrome patient are rare but with the advancement of the treatment, with increasing survival it appears that a growing number of these patients present with chronic renal failure.⁸

Patient with Down syndrome with multiple comorbidities with chronic renal disease undergoing major surgery with expected significant blood loss is very challenging for an anaesthesiologist to manage perioperative because we have to prevent hypotension in the same time, we have to restrict the fluid overload to prevent pulmonary oedema. So, in such cases volume to volume replacement with the colloid or blood and blood product with minimum crystalloid is the best management to avoid complications.⁹ In this case the patient is a down syndrome with hypothyroidism with CKD with anaemia was undergoing orthopaedic surgery in which significant blood loss was expected was managed with induction with titrated doses of propofol and intubation using atracurium. Maintenance

of anaesthesia was with O₂, air and sevoflurane. Blood loss was about 500ml which was managed with 1 unit (280 ml) of blood transfusion and 500 ml of crystalloids. Preoperative tranexamic acid was given to minimize blood loss.¹⁰

CONCLUSION

Down syndrome with chronic kidney disease undergoing major orthopaedic surgeries create major anaesthetic challenges due to expected significant blood loss, chances of fluid overload, multiple comorbidities and intellectual disability. Postoperative period also important as the patient may land up with complications. So a closed group management with orthopaedic surgeon, nephrologist, Anaesthesiologist, endocrinologist, nursing team is required to handle such complicated cases. A thorough preoperative evaluation in patients of Downs syndrome led to detection of kidney disease and hence effective closed group management was possible in this particular patient.

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CONFLICT OF INTEREST

The author declare there is no conflict of interest.

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