INTRODUCTION
Neurofibromatosis is the most common neurocutaneous syndrome. It is an autosomal dominant hereditary disease characterized by formation of neurofibromas all over the skin and various other body systems. The anaesthetic management of these patients requires attention to all possible abnormalities and associated disturbances to prevent any peri-operative complication. NF-1-associated complications of the musculoskeletal, respiratory, cardiovascular, central nervous, and gastrointestinal and genitourinary systems present with various degrees of considerations for anaesthesiologists. While evaluating and managing these patients for surgical procedures. NF-1 or von Recklinghausen disease, is an autosomal dominant disease with incidence of approximately 1/3000 births. Gastrointestinal (GI) neoplasms are present in about 2 - 25% of patients of NF-1. In these patients, neurofibromas are the most frequently (52%) diagnosed benign neoplasms. Patients with NF-1 and GI symptoms are at risk for gastrointestinal neoplasms; symptomatic patients are likely to experience significant morbidity. This report describes the anaesthetic of a patient with NF-1 and gastric outlet obstruction.

CASE REPORT
A 55 years female, known case of NF1, presented with history of recurrent episodes of vomiting for last 6 months, associated with abdominal distension especially after meals. These symptoms were progressively increasing. She did not have any surgery in the past. On examination, she was poorly built with multiple, cutaneous nodular neurofibromas present all over her body. Systemic examination revealed a normal cardiovascular and respiratory system. She was diagnosed as a case of gastric outlet obstruction and posted for surgery.

All her routine laboratory investigations were normal. Indirect laryngoscopy was done to rule out any lesion which may interfere with intubation and ventilation. Assessment of upper airway revealed no abnormality and indirect laryngoscopy was unremarkable. Surgery was planned under general anaesthesia. Informed written consent was obtained and anaesthetic procedure was explained to the patient. In the operating room (OR), intravenous access was obtained with 18 G cannula. Standard monitoring (ECG, SpO2 and non-invasive blood pressure) was instituted and baseline vitals were recorded. In OR, her blood pressure was 110/70, heart rate was 65 per minute and oxygen saturation 99%. Patient was considered as full stomach due to gastric outlet obstruction and rapid sequence induction was done. She was pre-oxygenated with 100% oxygen for 3 minutes. No positive pressure ventilation was given. Propofol and fentanyl were used for induction. Intubation was uneventful and was facilitated with rocuronium. Cuffed oral endotracheal tube of 7.5 mm size was used. Nasogastric tube was placed and gastric contents were aspirated. Anaesthesia was maintained with oxygen, nitrous oxide and isoflurane. The duration of the surgery was about 90 minutes. All hemodynamic parameters were stable during surgery and maintained within 20% of the base line values. Neostigmine and glycopyrolate was used to reverse the neuromuscular blockade after completion of surgery. Patient was extubated after return of airway reflexes and adequate muscle recovery. Postoperative period was uneventful. She was discharged on 6th postoperative day. Histological examination of the mural thickening in the pyloric antrum was found to be sub-mucosal neurofibromatosis.
NF-1 is a neurocutaneous syndrome with multisystem involvement. The diagnostic criteria for NF-1 include 2 or more of the following features: 6 or more café au lait spots (CALMs) > 0.5 cm in pre-pubertal patients or 1.5 cm or larger in postpubertal children or two or more of any type of neurofibroma or one or more plexiform neurofibroma, axillary or inguinal freckling, 2 or more lisch nodules of the iris, a distinct bony lesion, an optic nerve glioma, or a first degree relative with NF-1.2

NF-1 is a single-gene disorder, occurs due to heterozygous mutation of the NF-1 gene which results in loss of activity or in a non-functional neurofibromin protein. Neurofibromin’s function is not completely understood but it is known to be a guanosine triphosphatase - activating protein. Its functions is to downregulate p21-ras (rat sarcoma), a cellular proto-oncogene which is an important regulator of cell growth. So, neurofibromin has a role in the control of cell division and possesses tumour suppression properties. Pathologically, neurofibroma in NF-1 patients may be of the localized, plexiform, or diffuse type. Localized neurofibroma appears as non-encapsulated, well-defined tumour involving single nerve. Microscopically, it is formed by Schwann cells and fibroblasts with a myxoid or mucinous matrix surrounded by collagenous tissue with mast cell infiltration. Plexiform neurofibroma involves multiple nerve bundles or a plexus of nerve and has same histological features as localized neurofibroma except for higher cellularity and collagen. Diffuse neurofibroma is common in the head and neck region and presents as plaque-like lesions on the skin with characteristic presence of fat seen on microscopy. In gastrointestinal neurofibromatosis, mostly patients have localized or plexiform neurofibroma along with diffuse thickening of the myenteric plexus.

NF-1 exhibits complete penetrance with variable expressivity. Clinical manifestations are extremely variable even among patients with the same genotype.3 Therefore, a thorough evaluation of anticipated complications must be done while planning anaesthesia for such patients. A careful systemic assessment is done to decide the anaesthetic technique in patients with neurofibromatosis.

The examination includes assessment of airway, respiratory, cardiovascular and neurological systems along with examination of vertebral anomalies.

Intra-oral manifestation of NF-1 present in about 5% of the patients which includes neurofibroma of tongue, larynx, aryepiglottic folds or arytenoids, involving areas which are rich in terminal nerve plexuses.4 Hypertension is main cardiovascular manifestations of NF-1 and may be associated with pheochromocytoma or renal artery stenosis. Vasculopathy, cardiomyopathy, heart defects and superior vena cava obstruction are other cardiovascular complications. Pulmonary pathology includes pulmonary fibrosis and cystic lung disease.

There should be proper pre-operative assessment including questioning about cardiovascular disease, reviewing prior echocardiography reports and performing screen for hypertension. Nephrotoxic agents should be used cautiously. Pre-operative evaluation includes pulmonary function testing, indirect laryngoscopy to examine involvement of airway, computed tomography (CT) or MRI to investigate for any respiratory or neurological complication.5 In NF-1 patients with difficult airway, American Society of Anaesthesiologists guidelines for difficult airway is to be followed.

Radiographic examination of the neck should be done in patients with multiple cervical neurofibromas to avoid spinal cord damage and painless dislocation of cervical vertebra during laryngoscopy and tracheal intubation.6 Intra operatively close monitoring of heart rate and blood pressure should be there with high index of suspicion for the possibility of pheochromocytoma and carcinoid tumour.

These patients have high incidence of epilepsy, learning difficulties and undiagnosed CNS tumours. Involvement of brain stem structures by neurofibroma can result in central hypoventilation syndromes. These patients may exhibit difficulty in weaning from mechanical ventilation postoperatively.7 Gastrointestinal tumours in NF-1 present with disordered gut motility, abdominal pain, hematemesis and melena. Complications as obstruction and perforation can occur and may be first manifestation of neurofibromatosis.8 Genitourinary tract may also be involved and retroperitoneal neurofibroma may lead to ureteric obstruction and hydronephrosis. Bladder catheterization may be difficult because of outflow obstruction.4
In neurofibromatosis-1 patients, spinal cord neurofibromas should be ruled out by CT or MRI before epidural or spinal procedures are performed. The presence of scoliosis, kyphoscoliosis or spinal cord tumours can make the use of spinal or epidural anaesthesia challenging for an anaesthesiologist.

In patients undergoing neuraxial block a postoperative neurological examination is essential to ensure that the patient has returned to his or her pre-existing neurologic status.

This case highlights the importance of careful preoperative assessment and pre-emptive management of neurofibromatosis-1 patients during surgery. NF-1 is one of the most common genetically transmitted disease and we are likely to encounter such patients in our routine practise. Manifestations of patient with neurofibromatosis are often mild but there may be associated abnormalities of various systems highlighting the importance of anaesthetic management of patient with this disease. Patients with the disease require follow-up and management by multiple disciplines throughout the course of their lives. It is important to have working knowledge of the clinical manifestation of this disease. A systematic approach to the pre-operative assessment and a comprehensive anaesthetic plan for each patient of neurofibromatosis can result in better perioperative management.

REFERENCES