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ANAESTHESIA MANAGEMENT OF FEMUR FRACTURE FIXATION IN PATIENT WITH FANCONI SYNDROME – A CASE REPORT

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Abstract –

Detoni Debre Fanconi syndrome also called as Renal Fanconi syndrome is a rare acquired or inherited condition involving generalised transport defect in the proximal convoluted tubule of nephron leading to renal loss of electrolytes or substances that are usually absorbed. The incidence rate of this syndrome is 1:40,000 live births. Here we present the anaesthesia management in a patient with Fanconi syndrome posted for fixation of fracture femur.

Case Report –

A 22Year old female weighing 34kg, height of 140cm with history of Fanconi syndrome presented to our institution with Fracture femur due to trivial trauma. She underwent evaluation for short stature 10 years ago and was diagnosed with Fanconi syndrome and she has been on treatment. 2 years prior to the current complaint, she underwent plating for fracture left femur. Her parents had the history of consanguineous marriage and her sibling has similar complaints. On examination, she has short stature and bowing of both lower limbs. Her daily medications included Potassium chloride syrup 10ml, Sodium bicarbonate 500mg, Calcium, Phosphorous, Folic acid supplementation. Her preoperative Hemoglobin was 10.5g/dl, Arterial blood gas analysis showed pH – 7.23 ,serum sodium -136 mmol/l,serum potassium -2.7mmol/l (3.5-5.5 mmol/l) ,serum bicarbonate -13.4 mmol/l(23-30 mmol/l),Serum calcium – 0.80 mmol/L(2.2-2.7 mmol/l).Ultrasound abdomen showed Bilateral medullary nephrocalcinosis. Chest Xray and ECG were normal. Complete urine examination was normal. After supplementation with bicarbonate and potassium on the day before surgery, her Arterial blood gas analysis showed pH -7.29 , serum sodium -132 mmol/l, serum potassium -3.4mmol/l,serum bicarbonate – 16 mmol/l,Serum calcium- 1.08 mmol/l.

General anaesthesia was adminstered with titrated doses of anaesthetic agents and endotracheal intubation was done. Sevoflurane was given for maintenance. Intraoperative Arterial blood gas analysis showed –pH-7.26,serum sodium -138 mmol/l , serum potassium 2.8mmol/l,serum bicarbonate -16.2mmol/l , serum calcium -0.73mmol/l. Blood glucose was 90mg/dl. Intraoperatively 7.4%Sodium bicarbonate, Potassium chloride infusion was started and DNS was given for correction. Intraoperative vitals were stable. After the uneventful procedure of TENS nailing to right femur, the patient was extubated and shifted to the postoperative recovery unit.Post operative Arterial blood gas analysis showed - pH-7.23, serum sodium – 136 mmol/l,serum potassium 2.7mmol/l,serum bicarbonate -13.4mmol/l ,serum calcium -0.80 mmol/l. Post operatively she was closely monitored for ECG changes in view of low potassium in ABG despite intra operative correction and respiratory distress in view of acidosis . She was stable through out the post operative period and she was continued on her oral supplementation for electrolytes later.

Discussion -

Detoni Debre Fanconi syndrome or Fanconi renal tubular syndrome is a rare acquired or inherited condition involving the proximal tubule with the renal loss of glucose, phosphate, bicarbonate ,calcium, uric acid, amino acids. This disease presents an inherited form and acquired form. Inherited form occurs early age and Acquired form occurs at any age.Adults with Fanconi syndrome typically have the acquired type and children typically have the genetic type. Inherited form occurs in 1:40,000 live births. The defect of the gene is seen on 15q 15.3 1 .It follows an Autosomal Dominant pattern, but Autosomal Recessive form and X lined transmission have been reported.

Causes –

Inherited

Cystinosis
Galactosemia
Tyrosinemia
Hereditary fructose intolerance
Lowes syndrome
Mitochondrial disease

Acquired –

Drugs – cisplatin,ifosfamide,tenofovir ,adefovir2
Dysproteinemia – multiple myeloma, Sjogren's syndrome, Amyloidosis
Heavy metal poisoning-Lead, cadmium

There is no well-established pathophysiology for the disease. Probable pathologies are functional modifications in the carriers that transport ingredients across the luminal membrane, disorders in the cellular energy metabolism, changes in the permeability of the tubular membrane[3]

Clinical manifestations of Fanconi syndrome vary depending on the pathogenesis of the disease.Inherited form presents with proximal renal tubular acidosis ,hypophosphatemic rickets,hypokalemia,polyuria,polydipsia. Acquired form features slightly different abnormalities such as renal tubular acidosis,hypophosphatemia,hypokalemia ,osteomalacia and muscle weakness.[4]

The idiopathic Fanconi syndrome occurs in the absence of any perceptible cause, and most cases are sporadic. Most often it features recurrent episodes of dehydration, rickets, and failure to thrive.[4]

Prognosis of the patient depends on the cause of the syndrome and the severity of renal and extra renal manifestations.Genetic forms are difficult to manage .They are usually associated with disruption in the growth and involve other organs.4

Features of Fanconi Syndrome –

Metabolic abnormalities –

Glycosuria
Hyperaminoaciduria
Hypophosphatemia
Acidosis
Hypokalemia
Hypouricemia

Clinical features-

Rickets in children
Osteomalacia in adults
Growth retardation
Polyuria
Dehydration
Progressive renal failure

Patients are said to be anxious. Consequently, sedative premedication is recommended. Regular assessment and replacement of electrolytes must be done. Careful planning and attention to fluid, electrolyte management are indicated in the perioperative period. Extensive monitoring will facilitate a prompt, appropriate response to rapid and large volume shifts that may be encountered. Careful positioning and handling are required because of rickets and osteomalacia. Avoid drugs eliminated mainly by the renal system. Some drugs such as certain antibiotics or neuromuscular blockers may require dose adjustments. Scoline is not contraindicated . Hypoventilation should be avoided intraoperatively to prevent antecedent respiratory acidosis which can add upon the pre-existing metabolic acidosis. The patient is prone to hypoglycaemia, so blood glucose needs to be monitored at regular intervals. Arrhythmias can occur due to hypokalemia, and other electrolyte abnormalities, so vigilant monitoring is required. Hypophosphatemia and hypokalemia impair the contractility of the diaphragm and may lead to delayed weaning from mechanical ventilation.

Conclusion :

Fanconi`s syndrome is a rare congenital or acquired syndrome. It may precipitate many metabolic abnormalities which if not corrected adequately leads to respiratory and cardiovascular complications perioperatively. Utmost care is required during the positioning of the patient. vigilant monitoring of the patient for perioperative complications is crucial

Fig 1 – Short stature of the patient

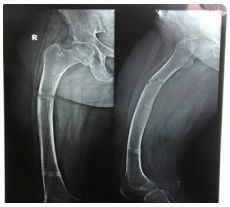

Fig 2 – Fracture of right femur

Fig 3 – Intra operative monitoring of vitals

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