CASE REPORT

Anaesthetic management of a case of osteogenesis imperfecta for lower limb surgery

- Dr Malathi .C.N[1]
- Dr. P.N Viswanathan[2]
- Dr Raghavendra Babu T[3]
- Dr. Kavya N[4]

SUMMARY

Anaesthesiologist are involved in paediatric emergencies through the administration of anaesthesia for emergency surgeries and have a role in airway management, resuscitation and intensive care.

Neonates have special risks associated with anaesthesia and surgery. They present great challenges to the anaesthesia team. Ideally surgery for neonates should be undertaken in a specialist unit. Neonatal defects which need emergency surgical care are congenital diaphragmatic hernia, omphalocele, gastroschisis, malrotation of gut, volvolus, intususception and others.

KEY WORDS :

Gastroschisis, anaesthetic management.

CASE REPORT :

A one day old baby posted for gastrochisis was delivered by LSCS. The mother had regular antenatal checkups. Soon after birth, intestinal contents along with sac present in the lateral aspect of umbilicus. Child was shifted to NICU care and was not in respiratory distress, maintaining SpO2 95% in room air, IV fluids 5% dextrose with calcium gluconate. General anaesthesia was planned for the reduction and closure of the defect.

ANAESTHETIC MANAGEMENT :

It consisted of intensive monitoring aides like ECG, blood pressure, oxygen saturation, temperature, end tidal CO2. Neonate was induced with use of N2O and O2 and with intravenous induction with pentothal sodium 2mg/kg, atropine 0.01mg/kg succinylcholine 1mg/kg and intubated with 3mm ETT , bilateral air entry equal and care was taken to prevent displacement. Maintainance with N2O, O2 + Atracurium 0.5mg/kg.Later neonate was put on post-op ventilatory support for 2 days and then weaned.

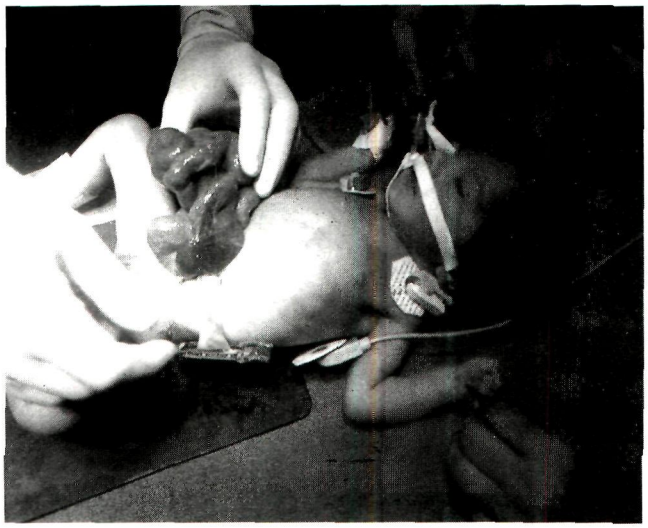
1 Associate professor, Department of Anaesthesiology, J.S.S Medical college and Hospital, Mysore -04.

2 Professor and HOD, Department of Anaesthesiology, J.S.S Medical college and Hospital, Mysore -04.

3 Post graduates- (MD), Department of Anaesthesiology, J.S.S Medical college and Hospital, Mysore -04.
Email- dr.babu77@gmail.com

4 Post graduates- (MD), Department of Anaesthesiology, J.S.S Medical college and Hospital, Mysore -04.

The neonate was shifted from NICU to the post operative ward.



DISCUSSION :

GASTROSCHISIS :

(Paraomphalocele, Laparoschisis, Abdominoschisis)

Derived from a Greek word ***GASTRO: STOMACH; SCHUSIS: TO SPLIT***

It's a congenital abdominal wall defect where all the layers of abdominal wall involved with evisceration of abdominal organs. Abdominal wall defects are diagnosed antenatally as early as 3rd trimester by amniocentesis and by ultrasound, and MRI scan. There is also a scope for foetal surgery if diagnosed in the midtrimester by hysterotomy and performing surgery on the foetus in utero with the placental circulation and oxygenation through the placenta. If diagnosed in the late last trimester, delivered by modified LSCS and surgery can be performed on the newborn. (EXIT).

Multifactorial, autosomal recessive type of inheritance. Obstruction of omphalomesentric vessel during development leads to ischemia.

INCIDENCE :

One in 3000 normal births, male to female ratio 3:2. There may be associated disorders with gastroschisis like intestinal stenosis (15%) intussusception, volvolus, malrotation, intestinal damage due to exposure to amniotic fluid. Anomalies like VSD, ASD; diaphragmatic hernia, anencephaly, scoliosis, cleft lip and palate, amniotic bands, may be present. IUGR is due to vascular accidents, loss of protein across the exposed bowel wall into the amniotic fluid and poor perfusion. Bowel wall is thickened with a fibrin peel due to exposure to amniotic fluid. It also may involve stomach, bladder, uterus and liver.

Some of the differential diagnoses are omphalocele, bladder extrophy, cloacal dystrophy, *limb body wall complex, Pentology of Cantrell.*

SURGICAL MANAGEMENT:

Gastroschisis requires urgent surgery as the bowels are exposed and fluid and heat loss may be significant. There is a risk of compromised gut circulation with ischemia and infarction. Abdominal compartment syndrome may occur if the abdominal contents are reduced under pressure when abdominal cavity is small.

Staged surgeries require application of silo to cover abdominal contents. The silo is suspended over the contents and reduced under gravity over 4-7 days, before definite abdominal closure.

Recently, a preformed siliastic spring loaded silo is inserted and a serial reduction is done in NICU without any anaesthesia or sedation and delayed closure is done when bowel is adequately reduced. This reduces the repeated exposure of baby for anaesthesia and does not require ventilatory support in NICU. It improves fascial closure rates, more rapid returning of bowel function and fewer complications compared to initial early repair. It would seem to be a technique ideally suited to use for treatment in developing world.

Post operative ↑ IAP

1. ↓ Organ perfusion leading to decreased intestinal, renal, hepatic perfusion, impaired organ function,

oedematous bowel, decreased urine output, decreased venous return.

2. ↓ Ventilatory reserve due to decreased diaphragmatic activity, patchy lower lobe pneumonia, atelectasis and respiratory failure.

Some babies with small defect can breathe spontaneously after surgery but majority require ventilation due to increased abdominal pressure with compromised respiratory function in NICU. The neonates may require muscle relaxants for 24hrs. Analgesia can be provided by fentanyl / morphine, caudal epidural, paracetamol 10mg/kg, 6th hrly IV/ PR. Regional technique- combined spinal epidural can be done for surgical correction and post-op pain relief.

TOTAL PARENTERAL NUTRITION : may be needed if paralytic ileus is expected and continued till full feeds are established. Dopamine administration may be necessary to treat hypovolemia, hypotension and maintain urine output. Some of early complications are necrotising enterocolitis, renal insufficiency pneumonia, cellulitis of abdominal wall, cholestasis, and gastroesophageal reflux can occur

Neonatal care begins in the NICU soon after the birth. It includes, care of bowel, intravascular volume expansion, assessment of respiratory distress, prevention of hypothermia etc.

PRE-OP ASSESSMENT:

1. **AIRWAY ASSESSMENT**: Difficult intubation in neonates may be associated with micrognathia or abnormal tissue (large tongue as in Beck-With Weideman). Elective intubation may be necessary for a transfer to a specialist centre.
2. **RESPIRATORY ASSESSMENT**: Is essential as respiratory compromise can occur with reduction of the abdominal content into the abdomen.

Post-op apnoea is more in preterm neonates with H/O preoperative apnoea and <44weeks of post conceptual age (PCA), babies with RDS, PDA or neurological disorders. Other causes like General anaesthetics, opioids, hypothermia, dehydration, anaemia. (HCT <30%).

3. **INTRA VASCULAR VOLUME STATUS**: Gastrochisis may present with significant fluid loss results in fluid resuscitation. The neonate is unable to compensate for dehydration and blood loss and therefore it is imperative to assess and correct the fluid deficit. They have poor control over the capacitance vessels and baroreceptor activity is immature, which leads to difficulty in maintaining cardiac output in the presence of hypovolemia.

Cardiac output is primarily rate dependent. Vagal tone predominates leading to bradycardia. The neonatal myocardium contains 30% less contractile tissue and more connective tissue and less complaint ventricle immature sympathetic system and stores of adrenaline are less. Hypoxia, acidosis, hypovolemia, and electrolyte imbalance leads to dysrhythmias.

4. **NERVOUS SYSTEM**: Functionally and anatomically immature, cerebral circulation-intraventricular haemorrhage can occur due to neonatal asphyxia, apnoea, osmotic effect of large doses of 8.4% sodium bi carbonate, use of 10% dextrose, hypertension caused by inadequate analgesia and stimulation by awake intubation.

TEMPERATURE CONTROL:

It is one of the most important considerations in anaesthetic management of the neonate. The neonate has increased metabolic requirement decreased body reserve of carbohydrates, decreased liver function, and increased chances of hypoglycaemia .Hypothermia causes metabolic acidosis, left to right shunt respiratory depression and hypoventilation, delayed recovery from anaesthesia, prolonged action of drugs, difficulty in assessing hypovolemia, dysrrhythmia and cardiac depression.

FLUID MANAGEMENT :

Goals of perioperative fluid administration: Maintain tissue perfusion and normoglycaemia, treat pre-existing water and electrolyte disturbance if any and avoid creating new disturbances by such therapy

Total intraoperative fluid = compensatory intravascular volume expansion+ deficit + maintainance + loss + third space loss

Maintainance – 4: 2: 1 formula

5 % D with ½ saline + 20 meq/lt K^+

¼ saline + 20 meq/lt K^+ in neonates due to decreased ability to handle sodium load

Deficit : Wt * 4 ml/kg* hr

50% deficit in 1st hr

25% of deficit in 2nd hr

25% of deficit in 3rd hr

Preoperative deficits are replaced with balanced salt solution or 1/3 NS Colloids can be used to replace third space loss (gelatin, starch, and albumin). Isotonic solution is used to correct hypovolemia.

TRANSPORTATION AND MAINTAINING NORMOTHERMIA :

Neonates may develop apnoeic episodes, bradycardia and desaturation during transportation from NICU to OR. This occurs due to excessive handling, hypoxemia, and airway obstruction. So it is important to monitor heart rate, respiration, O_2 saturation. Preferably transported in incubator with O_2 and resuscitation drugs and equipments. Normothermia can be maintained by air heating and humidification, cutaneous warming by use of surgical drapes, cotton blankets, plastic bags. They reduce the heat loss by 30%. IV fluids administered at ambient temperature.

CONCLUSION:

Neonates with abdominal wall defects require particular attention to

1. Fluid resuscitation and temperature control
2. Attempts by the surgeon to close the defect causes a decrease in blood pressure, cardiac output and may interfere with ventilation.
3. A silo can be placed with a delayed closure.
4. Amount of insensible loss is inversely proportional to the gestational age
5. Neonatal kidney is unable to excrete large amounts of excess water
6. Every child should be assessed for their own status of volume, fluid and electrolyte and plan as per the need at that time

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