AFRICAN JOURNAL OF MEDICINE and medical sciences

VOLUME 24, NUMBER 4, DECEMBER 1995

EDITOR: B.O. ONADEKO
ASSISTANT EDITORS:
B.O. OSOTIMEHIN and A.O. UWAIFO

SPECTRUM BOOKS LIMITED
Ibadan • Owerri • Kaduna • Lagos

ISSN 1116–4077

Afr. J. Med. med. Sci. (1995) **24** *403-406*

Anaesthetic considerations in exomphalos repair: a report of 2 cases

A.O. AMATA
Department of Anaesthesia Usmanu Danfodiyo University Teaching Hospital, Sokoto, Nigeria.

Summary

The anaesthetic management of two neonates with exomphalos associated syndromes for surgical repair are described. The technical advancements in the surgical and anaesthetic management of these neonates which have resulted in reduction in morbidity and mortality are discussed. The high incidence of associated anomalies and the problems they pose to the anaesthetist in addition to the usual challenges of neonatal anaesthesia are highlighted.

Résumé

Ceci est une déscription du traitement anesthétique de deux nouveaux-nés avec des syndromes relatifs à l'exomphale lors des réparations chirurgicales. Les progrès techniques dans le maniement chirurgical et anésthetique de ces néonates qui a reduit la morbidité et la mortalité sont discutés.

L'incidene élévée des anomalies associées et les véritables gageures qui se présentent naturellement en anesthésie néonatale sont mis en vedette.

Introduction

Exomphalos or omphalocele is a congenital abdominal wall defect. A number of syndromes are commoly observed in neonates with exomphalos [1,2]. Apart from the anaesthetic problems associated with surgical repair of an exomphalos in a newborn, the presence of these syndromes and associated congenital anomalies usually have significant anaesthetic implications [2, 3]. There has been a paucity of information in the literature relating to the anaesthetic management of these neonates despite their obvious anaesthetic significance. Two illustrative cases of exomphalos associated syndromes are presented to highlight the challenges and problems these conditions may pose to the anaesthetist.

Case Reports

Case 1

A one-day-old, 4.8 kg male neonate and second child of a 20-year-old Nigerian mother delivered at full term by spontaneous vertex delivery was referred to the paediatric surgical clinic because of the presence of an exomphalos. On examination he was a big plump neonate with obvious macroglossia, low set ears and a prominent occiput. The heart rate was 135 beats per min and regular. The heart sounds were normal and there was no murmur. The respiratory rate was 24 breaths per min and the chest was clinically clear. There was an exomphalos with an intact sac which measured 9cm in diameter. Haemoglobin concentration was 18gm. dl^{-1} serum urea 4.2 $mmol.L^{-1}$ (normal 2.5 — 6.5 $mmol.L^{-1}$). Blood glucose estimation revealed severe hypoglycaemia of 1.1 $mmol.L^{-1}$. The intravenous urography ((IVU) was normal. In view of the presence of exomphalos, macroglossia, gigantism and hypoglycaemia, a diagnosis of Beckwith — Wiedeman syndrome was made and the patient was scheduled for surgical repair of the exomphalos.

An intravenous infusion of 5 percent dextrose solution was set up and a bolus injection of 10 percent dextrose was given to correct the hypoglycaemia. A nasogastric tube was inserted and left on free drainage with intermittent frequent aspirations.

On arrival in the operating room, the patient was placed on a thermostatically controlled heated mattress maintained at 37°C. Atropine 0.02mg kg^{-1} iv was given immediately prior to induction of anaesthesia which was inhalational using oxygen,

nitrous oxide and halothane delivered via the Jackson Rees modification of the Ayre's T-piece circuit.

After an adequate depth of anaesthesia had been achieved, the patient's trachea was intubated with a 3.5 ID plain portex orotracheal tube and anaesthesia was maintained by controlled ventilation with the same agents. The patient was monitored with a precordial stethoscope and an automatic non-invasive blood pressure recorder (EME London). The exomphalos was surgically corrected by primary closure. The blood glucose level was monitored intra-operatively with a dextrostix. Blood loss was 60mls and this was replaced with crystalloid fluid. At the end of the surgical procedure the patient was extubated when he was awake. He was then transferred to the Neonatal Intensive Care Unit for close monitoring during the first 24 hours post-operatively. Blood glucose was estimated 2 hourly and glucose infusion was regulated as found necessary. The post-operative period was uneventful and the patient was discharged home on the ninth post-operative day.

Case 2

A one-day-old male neonate weighing 1.98kg was referred from the labour ward to the paediatric surgeon because of the presence of a major exomphalos. He was born prematurely at 36 weeks of gestation and had to be actively resuscitated by the doctor and midwife who assisted in the delivery. On examination he was noted to be a preterm, small for gestational age (SGA) infant. The heart rate was 168 beats per min, the heart sounds were heard and there was a systolic murmur. The lung fields were clinically clear. There was an exomphalos major consisting of a thin translucent sac with extra abdominal liver, spleen, small and large intestines and part of the stomach. The anterior abdominal wall was poorly developed. The packed cell volume (PCV) was 52%. Serum urea and electrolytes and blood glucose levels were within normal limits. A staged repair with application of prosthesis was planned. An orogastric tube was inserted for frequent aspiration and a secure venous access was provided by means of a saphenous vein "cut down" The patient was nursed in an isolette with overhead radiant heater preoperatively and transferred to the operating theatre in the isolatte.

He was placed on a thermostatically controlled electric mattress maintained at 37°C, atropine 0.02mg.kg^{-1} iv was given and anaesthesia was induced inhalationally with halothane, oxygen and nitrous oxide. Many attempts at tracheal intubation were unsuccessful. The surgeon had to infiltrate the edge of the sac with local anaesthetic (lignocaine) in order to suture the improvised silastic sac in place while the patient was breathing 50% nitrous oxide in oxygen (entonox) via a Rendell Baker face mask and Ayre's T-piece circuit. Monitoring was by a precordial stethoscope while fluid therapy was with 4.3% dextrose in 0.18% saline solution. The procedure lasted 25mins and the patient was transferred to the neonatal intensive care unit. The paediatrician and anaesthetist were involved in the post-operative fluid and nutritional management of the patient. However, the patient later deteriorated and died 48 hours after surgery. A postmortem diagnosis of Pentalogy of Cantrell was made based on the autopsy confirmation of the presence of a diaphragmatic hernia and sternal cleft in addition to an exomphalos and intracardiac cushion defects.

Discussion

The diagnosis of exomphalos is quite straightforward and is usually obvious immediately after delivery, therefore these neonates are presented for surgical correction within the first 24 to 48 hours [2]. The first 48 hours is the most vulnerable phase in the neonatal period and anaesthetic management is the most challenging and stressful to the anaesthetist [4, 5].

Ambrose Pare in 1634, was the first to describe an infant with exomphalos [6] and the first successful repair of a ruptured exomphalos was accomplished in 1873 by Visick using a skin closure [7]. The reported incidence varies between 1 in 3,200 and 1 in 10,000 live births [1,2,8,9]. The size of the anterior wall defect is variable and can range from 4cm to greater than 10cm. The sac containing the herniated viscera may contain small and large intestines, stomach, liver, spleen, bladder, uterus and ovaries.

Associated congenital anomalies are commonly seen in infants with exomphalos; these include abnormalities of the cardiovascular, genito-urinary, gastro-intestinal, musculo-skeletal and central nervous systems. The overall reported incidence of these severe associated anomalies ranges from 37% to greater than 75% [1,2,8,9].

There are a number of syndromes that are commonly observed in neonates with exomphalos

[1]. These include Beckwith—Wiedemann syndrome, Pentalogy of Cantrell, lower midline syndrome, and chromosomal syndromes.

In view of the high incidence of associated anomalies, it is imperative for the anaesthetist to perform a careful and thorough preoperative evaluation of the neonate. Of the many metabolic problems known to exist in these neonates, hypoglycaemia is the most common [3,10]. This is especially so in patients with the Beckwith — Wiedemann syndrome in whom it must be actively sought for. Adeyokunnu and Adeniyi [11] reported a 50% incidence of severe hypoglycaemia in Nigerian infants with this syndrome and it was the greatest contributor to mortality in their series. Maternal diabetes or abnormal glucose tolerance test was not found in any of the affected cases [11]. The hypoglycaemia is thought to be due to hyperinsulinism as a result of hyperplasia of the pancreatic islet cell [1]. Hypoglycaemia must be corrected pre-operatively. A continuous infusion of glucose should be maintained throughout the perioperative period to avoid the potential cerebral insult due to hypoglycaemia. In cases of hypoglycaemia resistant to correction, 1 in 200 epinephrine and diazoxide to suppress insulin release has been shown to be effective [10].

Hypocalcaemia is also common especially in the preterm infant and requires preanaesthetic correction [5]. Thirty-three percent of neonates with exomphalos are preterm [2]. Differential diagnosis of the Beckwith — Wiedemann syndrome include congenital hypothyroidism and Down syndrome because of similar features like low set ears, hypertelorism, macroglossia and umbilical hernia [3, 12]. However, congenital hypothyroidism and Downs syndrome can occur in association with the B-W syndrome and this has been reported in the literature [8, 12].

Venous access is more difficult in neonates. Gigantism is a feature of the B-W syndrome. The above neonate weighed 4.8kg and was very plump. The average weight of a Nigerian neonate born at term is 3.0 kg [11]. Venepuncture was difficult in this plump baby because of the large amount of adipose tissue which made visualisation of peripheral veins impossible while in the preterm neonate a venous cut-down was necessary because the peripheral veins visible were very tiny.

Control and expert management of the airway is essential for the safe and effective administration of general anaesthesia. Various reports and reviews have highlighted the greater difficulty of tracheal intubation in the neonate because of its peculiar anatomical features [13, 14, 15]. While many of these cases of "problem intubation" can be anticipated with adequate pre-operative evaluation, occasional cases arise unpredictably at laryngoscopy. Tracheal intubation may be very difficult in the B-W syndrome because of macroglossia, a large epiglottis, cephalad glottis and a prominent occiput. A large tongue makes insertion and manoeuvring of the laryngoscope difficult while a prominent occiput may limit the ability to adequately position the head on the neck. The small size of the preterm or SGA neonate with the proportionately small mouth may make the insertion of the conventional paediatric laryngoscope difficult and leave little or no room for manipulation. An anteriorly placed larynx increases this difficulty. Because of the anticipated difficulty envisaged in these patients the spontaneous inhalational technique was used for induction and intubation performed without the use of a muscle relaxant. While the first case was intubated without much difficulty, it was impossible to intubate the second neonate for the above stated reasons i.e. a small oral cavity with a high anterior larynx. He was therefore managed by tissue infiltration with a local anaesthetic, supplemented with inhalation of entonox via a facemask. A laryngeal mask airway (LMA) if available, would have been an effective alternative method of securing a patent airway in this patient. LMAs have revolutionized difficult intubation being extremely useful in securing airways in patients of all ages who are difficult to intubate but have adequate mouth opening. Intermittent positive pressure ventilation is also very effective with a LMA.

The patient with exomphalos is very prone to hypothermia, especially if the sac has ruptured, because of evaporative loss from the exposed viscera in addition to the fact that the neonate has a large surface area to body weight ratio and an inability to generate heat by normal mechanisms [1,13]. Hypothermia in these neonates must be vigorously avoided. Patients with ruptured exomphalos may present with shock, acidosis, hypovolaemia and secondary infection and may require aggresive preoperative fluid and electrolyte resuscitation and urgent operative surgery.

For most neonates with small to moderate exomphalos, primary and abdominal wall closure as a one stage procedure is feasible and is the preferred method of closure as was done in the first case. But

in large exomphalos respiratory function and venous return may be compromised by a tight closure [1, 13, 16]. In these patients, stage repair is usually done. This is by the application of a silastic or teflon prosthesis as suggested by Schuster [17]. Gradual manual reduction of the prosthesis is then performed. Post operative management will depend on the type of operative procedure performed. The neonate must be managed in a neonatal intensive care facility with trained personnel. If there is any respiratory impairment or compromise, the neonate must have ventilatory support with an infant mechanical ventilator [1,2]. This is more likely in cases of a tight primary closure. Although patients with an intact exomphalos usually have an early return of gastro-intestinal function and can resume enteral intake, neonates with ruptured exomphalos or those with a synthetic prosthesis usually have a prolonged adynamic ileus following repair. One of the major post operative improvements in the management of these patients has been the recognition of this problem and the application of parenteral nutritional therapy. The combined mortality rate from various reports is 36.4 percent with a survival rate of 63.3 percent [1]. The most important determinants of survival in patients with exomphalos were the size of the defect, the birth weight and the presence and severity of associated anomalies [1, 2, 9, 18].

References

1. Grosfeld JL, Weber TR. Congenital abdominal wall defects gastroshisis: and omphalocele In: Current problems in surgery. Year Book medical Publishers Inc. 1982:19.
2. Stoelting RK, Dierdorf SF, McCammon RL. Anaesthesia and coexisting diseases. Churchill Livingstone 1988: p. 832.
3. Naguib M, Redwan A, Khawaja S. Anaesthetic considerations in Beckwith—Wiedemann syndrome. MEJ Anaesth. 1987; 9:127-133.
4. Crone RK, Sorenson GK, Orr RJ. Anaesthesia for the neonate. Can. J. Anaesth. 1991; 38:R105-155.
5. Waugh R, Johnson G. Current considerations in neonatal anaesthesia. Can. Anaesth. Soc. J. 1984; 31:700-709.
6. Pare A. The works of that famous chirurgeon, Book 24. London: Th. Cotes and R Young 1634:59.
7. Visick C. An umbilical hernia in a newly born Child. Lancet 1873; 1: 829.
8. Allen RG. Omphalocele and gastroschisis. In: Paediatric surgery. Holder TM, Ashcraft KW (eds) Philadelphia. WB Saunders Co. 1980: 572-588.
9. Robertson NC. A manual of Neonatal Intensive Care. Edward Arnold Ltd. London 1986:276.
10. Schiff D, Coke E. Wells D, Stern D. Metabolic aspects of the Beckwith—Wiedemann Syndrome. J. Pediatr. 1973; 82:258- 261.
11. Adeyokunnu A, Adeniyi A. The Beckwith — Wiedemann syndrome in Nigerian infants E. Afr. Med. J. 1981; 58:684-490.
12. Martinez R, Ocampo — Capos R, Perez Aroyo R, Corona Rivera E, Cantu J. The Beckwith — Wiedemann syndrome in four sibs including one with associated congenital hypothyroidism. Eur. J. Pediatr. 1985; 143: 233-236.
13. Palahniuk RJ. Anaesthesia for neonatal surgical emergencies. Can. J. Anaesth. 1989; 36: 549-552.
14. Geffin B. Anaesthesia and the "problem upper airway". Int. Anaesth. Clinics. 1990; 28: 106-114.
15. Brown TK. Safe pediatric anaesthesia. Can. Anaesth. Soc. J. 1985; 32: 522-523.
16. Yaster M, Buck JR, Dudgeon DL *et al.* Hemodynamic effects of primary closure of omphalocele gasroschisis in human newborns. Anaesthesiology 1988; 69: 84-88.
17. Schuster SR. A new method for the staged repair of large omphalocele. Surg. Gynecol. Obstet. 1967; 125:837-839.
18. Evans J. Exomphalos and Gastroschisis, a study of fetal and neonatal outcome. J. Obst. Gynae. 1991; 11: 15-18.

(Accepted 29 June, 1993)