Challenges in the Anaesthetic Management of Congenital Diaphragmatic Hernia in a Tertiary Care Children Hospital of Nepal

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ABSTRACT

INTRODUCTION: Congenital diaphragmatic hernias (CDH) are complex developmental anomaly. It results from anomalous closure of pericardio-peritoneal canal. Consequently, abdominal organs extrude into the thoracic cavity, impairing the growth of the ipsilateral lung. Surgical correction is the only treatment modality. There are various challenges faced by anesthesiologists such as hypoxia and hypercarbia leading to pulmonary hypertension and right to left shunt.

This study is carried out to present perioperative complications that we faced during the anaesthetic management of CDH in patients who underwent surgical closure of the diaphragmatic defect in a tertiary care centre of Nepal.

METHOD: This is a retrospective study carried over a period of one year. We reviewed the medical records of children with the diagnosis of CDH who underwent surgical correction of diaphragmatic defect. The perioperative complications were recorded as the appearance of bradycardia, hypoxia, sepsis, DIC, pneumothorax and the collected data were analyzed.

RESULT: Thirteen children presented to the hospital with the diagnosis of CDH and underwent surgical repair. Among them, 9 (69.23%) were male and 4 (30.76%) were female. Hospital presentation on 8 to 30 days of life was noted in 46.15% of the cases, The most common complication during the perioperative period was hypoxia, bradycardia and pneumothorax. The survival rate was 61.53%.

CONCLUSION: The anaesthetic management of CDH is still a difficult and challenging for anaesthesiologist. Bradycardia, hypoxia, pneumothorax, septicemia and DIC are the major causes of perioperative morbidity and mortality. Preoperative optimization and gentle ventilation strategy makes significant impact on survival in child.

KEY WORDS: Congenital diaphragmatic hernia, hypoxia, pulmonary hypertension.

INTRODUCTION:

Congenital diaphragmatic hernia (CDH) is a complex developmental anomaly. It results from anomalous closure of pericardio-peritoneal canal. Consequently, abdominal contents extrude into thoracic cavity, impairing the development of ipsilateral the lung.

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The incidence of congenital diaphragmatic hernia in western population is 1 in 2000–5000 live births.[1] The left sided CDH is more common (85%) and right sided CDH are rare (10-15%).The etiology of congenital diaphragmatic hernia is unknown, however, 2% of cases have been noted to be familial and another 15% of patients have associated chromosomal abnormalities.[2]

CDH results from anomalous closure of pericardio peritoneal canal.[3] Consequently, abdominal organs (i.e. Stomach, parts of the descending colon, the left kidney, and the left lobe of the liver) develop in the pleural cavity which impaires the normal growth and development of ipsilateral lung and mediastinal shift contributes for hypoplasia of contralateral lung.[4]

Surgical repair of the diaphragmatic Hernia is the only treatment modality. In poor resource settings like ours, the challenges for the anaesthetic management of CDH are more as there are inadequate facilities for prenatal diagnosis, transportation and neonatal intensive care. Affected neonates usually present to the hospital late with respiratory distress and cyanosis. During perioperative period, various challenges are faced by anesthesiologist. They are acidosis, hypoxia, pneumothorax, sepsis and hypercarbia and these abnormalities are the predictor of survival of the child.[5] The perioperative mortality of children undergoing surgical correction of CDH is up to 50 to 60%.[6] The incidence and mortality rate of CDH in Nepali is not known.

We conducted this study to know the perioperative anaesthetics complications that were encountered during an elective surgical repair of CDH in the tertiary care children hospital of Nepal.

METHOD

This is a retrospective hospital based study carried out in Kanti Children Hospital of Nepal over the period of one year. We had reviewed the medical records of children diagnosed as CDH who underwent surgical correction of diaphragmatic defect from 2013 May to 2014 May. Ethical approval was obtained from the Hospital.

The clinical profile of CDH (i.e. age of children, gestational age, sex and clinical features, results of radiological investigations and presence of associated malformations at the time of presentation), operative findings, and outcome of treatment, follow-up status were entered into a recorded predesigned proforma. We also evaluated the survival rate and perioperative anaesthetic events and analyzed them. Data were recorded up to the postoperative period until discharge from hospital or death.

The perioperative complications were recorded as the appearance of bradycardia, hypoxia, sepsis and/or DIC (Disseminated intavarcular Coagulation). Bradycardia was defined as when heart rate decreased to below 60 beats per minute. Hypoxia was defined as decrease in saturation (<85%). Similarly sepsis was defined as presence of clinical sign and symptoms of SIRS in the presence of or as a result of suspected or proven infection.[7]

DIC was diagnosed on the basis of prolongation of the PT, reductions in the platelet count, and an increase in fibrinogen degradation products.

Classic triad of CDH includes; cyanosis, dyspnea, and apparent dextrocardia. For the purpose of this study, diagnosis of CDH was confirmed on the basis of clinical presentation (i.e. respiratory distress, respiratory tract infection, feed refusal, bulged chest, decreased or absent breath sounds on the left side, The heart sounds displaced to the right side, scaphoid abdomen and bowel sounds heard on the chest and gas filled bowel with mediastinal shift in chest radiograph.[8]

The demographic profiles, peri-operative anaesthetic events of all children were recorded and the collected data were analyzed by means of statistical software Statistical Package for the Social Sciences (SPSS) version 20. Analyzed data were presented in the form of tables. Age of the patients was expressed in ranges, rest of the parameter were expressed in percentage.

RESULT

Thirteen children were found with the diagnosis of CDH who underwent surgical repair during the defined study period. Among them, 9 (69.23%) were male and 4 (30.76%) were female with male to female ratio is 2.25:1. In this study 7.69% cases presented within 24 hrs of birth while 46.15% of the children presented to the hospital on 8th to 30th days of birth. (Figure -1)

These children were found to be presented with symptoms of respiratory distress, 69.23% followed by respiratory tract infection 23.07% and feed refusal in 7.69%. All the cases had left posterolateral defect and direct closure was done in all of them.

All the children were found to be optimized preoperatively with supportive care until the children were stable and significant comorbodities had been diagnosed and optimized. All the children were found to be anesthetized with general anaesthesia using endotracheal intubation.

The complications encountered during intraoperative period were bradycardia, hypoxia. Bradycardia was found to be responding well to injection atropine, and decrease oxygen saturation was corrected with gentle positive pressure ventilation.

Pneumothorax, DIC and sepsis occurred in post operative period.

Five children were died in post operative period. The overall mortality of CDH during perioperative period was 38.46%.

The survival rate was found to be 61.53%. It was seen better in the age group 1 month to 12 months.

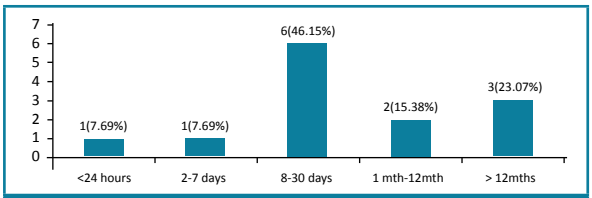

Fig 1: Age distribution of CDH presentation at hospital

Table 1: Perioperative Complications	
Perioperative events	Number of patients (%)
At induction	
Bradycardia	3 (23.07)
Hypoxia (<85%)	1(7.69)
Intraoperative	
Bradycardia	2(15.38)
Hypoxia (<85%)	1(7.69)
Post operative	
Pneumothorax	3(23.07)
Sepsis	3(23.07)
DIC	2(15.38)

Table 2: Age wise distribution of survival after surgery

Serial no	Age of neonates	No of Children Survived (%)	No of Children Expired (%)
1	<24 hours (n= 1)	0 (0)	1(100)
2	2-7 days (n= 1)	1(100)	0 (0)
3	8-30 days (n= 4)	2 (50)	2 (50)
4	1month-12 month (n= 5)	3 (60)	2 (40)
5	>12 months (n= 2)	2(100)	0 (0)

DISCUSSION

Kanti children hospital is the only paediatric tertiary care government hospital of Nepal. Thirteen children presented to the hospital with the diagnosis of CDH who underwent surgical repair over a period of one year. The less number of CDH in our study might be due to the inadequate prenatal diagnosis facilities and transportation. Most of the severe forms of CDH might had died in the immediate postnatal period before reaching the hospital.

CDH is a life-threatening condition that is not just anatomical defects of the diaphragm, but represent a complex set of physiologic derangements of the lung, the pulmonary vasculature, and related structures. Before 1980s, it is considered as a surgical emergency and correction of defect is carried out as early as possible. Now the concept has changed from performing emergency repair to delaying repair for at least 24–48 hours to allow for clinical stabilization.[9, 10] The rationale of delaying surgery is to allow maximum expansion of lung.[11] In our study all the children were optimized preoperatively in surgical paediatric intensive care unit then went for the surgical correction of diaphragmatic defect.

CDH is found predominant in male child in our study with the male female ratio of 2.25:1. Similar findings were observed by Torfs et al, Khemakhem et al and Yang et al in their study.[12,13, 14]

In our study, six children (46.15%) presented to hospital on 8th to 30th days of birth. All the children were found to be referred from the different parts of Nepal. So the children might have presented late. In a study done by Jain et al [15] also noted that maximum cases were presented at 8-30 days of life. Abubakar et al reported the same finding in their study.[11]

In this study, regarding the clinical presentation, maximum number of children 69.23%, presented with complains of tachypnoea followed by respiratory tract infection 23.07% and feed refusal (7.69%). Similar findings were reported by Robinson and Fitzgerald in their study.[16]Another study carried out by Mei-Zahav M found that respiratory symptoms (43%), gastrointestinal symptoms (33%), both respiratory and gastrointestinal symptoms (13%), and asymptomatic (11%) in their study.[17] The findings of our study are consistent with them.

In our study, Left posterolateral defect was found in all cases of CDH. Gudbjartsson T et al , Colvin et al reported that the left side CDH is more common (69-87%) in their study.[18,19] But Mullins et al reported that sixty-eight percent (68%) of CDH was found on the right side, 18% on the left side, and 14% occur bilaterally which was different compared to other studies.[20]

The complications encountered during intraoperative period were bradycardia and decrease oxygen saturation. These Challenges faced by us during intraoperative might be due to the reversal of shunt in the presence of already existent pulmonary hypoplasia, pulmonary hypertension, and systemic hypotension due to mediastinal shift. The primary pulmonary hypoplasia and pulmonary hypertension would lead to hypoxemia. Therefore, in this study all child were ventilation with low tidal volume and airway pressure was maintained at <25 cmH2O during the intraoperative period to prevent hypoxemia. In spite of that one child developed hypoxia during induction and one developed during intraoperative period, manage with positive pressure ventillation. The pneumothorax, DIC and sepsis were found in postoperative period. Causes of pneumothorax can be associated with barotrauma. Children were kept low inspiratory pressure (<25 cmH2O) and low tidal volumes in postopetative ventilator setting in spite of that 3 children (23.07 %) developed pneumothorax.[21]

In our study five children were expired in post operative period. Three children expired due to sepsis and two children expired because of DIC. The overall mortality of CDH during perioperative period was 38.46%. In Lochbuhler H study, thirty newborns were anesthetized for repair of a CDH and reported the overall mortality of 27%.[22]

In our study the survival rate is 61.53%. Survival rate was better in the age group 1 month to 12 months. Different survival rates have been found in different studies. Sigmund H Ein and collegue found 36.9% of survival,[23] Langer et al have reported a survival of 50%.[24] We could not find the incidence and survival rate of CDH in Nepali literature. In Indian literature, Anurag Krishna and collegue reported 57%[25] survival rate. Raghvendran et al found an overall survival rate of 62%.[26] CDH has continued to pose a significant challenge to anaesthesiologist. Despite advances in prenatal diagnosis, transportation, paediatric intensive care unit (PICU) care and new treatment modalities,(i.e. High Frequency Oscillatory Ventilation, Extra Corporeal Membrane Oxygenation and inhaled Nitric Oxide) overall mortality rate has remains sill high.[27] But developing country like Nepal, lack all these neonatal advanced care facilities, as well as there are no adequate facilities for prenatal diagnosis and transportation, the challenges for the management of CDH are more. In our set up most of the severe cases of CDH were died before they reach to the hospital and only a few number of children with less severe pulmonary hypoplasia were survive. So we found better result in this study and we cannot compare our results with these researcher.

CONCLUSION

CDH is a complex developmental anomaly of children. Surgical correction is the only treatment. The anaesthetic management is still a difficult and challenging for anaesthesiologist in a low resource country. Bradycardia, hypoxia are common intraoperative problems. pneumothorax, septicemia and DIC are causes of postoperative morbidity and mortality. Preoperative optimization and gentle ventilation strategy with low tidal volume, airway pressure <25 cmH2O makes most significant impact on survival in child with CDH.

In poor resource settings like ours, the anaesthetic management of CDH is still challenging as there are no adequate facilities for prenatal diagnosis, transport and neonatal intensive care.

A large cohort study is required to estimate the incidence of CDH, frequency of various perioperative complications, causes of mortality and survival rate following surgical repair of CDH.

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