
Congenital Diaphragmatic Hernia Outcomes in East Africa: The Ethiopian Experience

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Background: *Despite advances in care leading to improved survival rate in high-income countries, congenital diaphragmatic hernia (CDH) continues to have a poor prognosis in sub-Saharan Africa. This retrospective analysis documents the demographics, presenting symptoms, initial diagnosis and outcomes of those CDH patients on whom operations are performed at TikurAnbessa Specialized Hospital (TASH), Ethiopia's largest tertiary referral center, from September 2012 to August 2016.*

Methods: *The pediatric surgery database was reviewed for those patients who underwent CDH repair, and these cases were retrospectively analyzed. All work was performed in compliance with the Addis Ababa University institutional review board.*

Results: *Out of 15 patients who underwent operations, twelve cases were included in our study. Average age at presentation to TASH was 233 days. 83.3% of our study patients were initially misdiagnosed; 50% were initially diagnosed with pneumonia. The diaphragmatic defect was on the left in six (50%) of our patients and on the right in six (50%). Two patients died after surgery. The remaining ten survived. All patients underwent primary repair via lateral subcostal incision. Average length of stay was 24.5 days.*

Conclusion: *Misdiagnosis of CDH remains to be a major problem in sub-Saharan Africa, likely contributing to delay in diagnosis and early appropriate care. First line physicians and neonatal care units should consider the possible diagnosis of CDH when neonates and infants present with respiratory symptoms.*

Key words: Congenital, diaphragmatic, hernia**DOI:** <http://dx.doi.org/10.4314/ecajs.v21i3.2>**Introduction**

Congenital diaphragmatic hernia (CDH) remains one of the major diagnostic and management challenges of pediatric surgery. How, when and if to operate on these patients are all hotly debated topics across the scientific and medical literature. It is presumed that one third of patients affected by CDH die in utero and one third will die in the neonatal period. Indeed, up to 30% of infants who have symptoms within in the first 6 hours of life will die¹. Those who present later in life (>24-36 hours after birth) have a presumed survival approaching 100% with appropriate care at a well-equipped tertiary referral center². In more severe cases, surgical intervention can be postponed until the patient is stable from a cardiorespiratory standpoint; survival is often more dependent on management of physiologic sequelae of CDH, including pulmonary hypertension and respiratory distress, rather than on repair of the hernia itself. That is why CDH is more of a physiologic than surgical emergency. Advances in neonatal care have improved survival of early presenters in high income countries, but prognosis remains poor in Africa³.

Tikur Anbessa Specialized Hospital (TASH) is Ethiopia's largest tertiary referral center and the only pediatric surgical referral hospital in the country. There are currently 5 full time pediatric surgeons and three pediatric surgery fellows. TASH serves as the training base for over 150 general surgery and other residents as well as medical students. Cases have been collected into a database of 6900 general pediatric surgical patients who have undergone operations between September 2012 and August 2016. Patients who underwent surgery for CDH (0.21%) were identified from this database. This retrospective analysis seeks to document the demographics, presenting symptoms, initial diagnosis and outcomes of those cases.

Patients and Methods

The TASH pediatric surgery database of cases from September 2012-August 2016 was reviewed for those patients who underwent congenital diaphragmatic hernia repair. Cases were included if charts could be located for review. Fifteen patients underwent operations. 12 cases were ultimately included in the study. All work was performed in compliance with the Institutional Review Board at TASH.

Results

Fifteen patients underwent surgical repair at TASH from September 2012-August 2016. Three of these patients were excluded from this study because their hospital charts could not be located. The remaining 12 (80%) formed the basis of our analysis (Table 1). Their ages ranged from 10 hours to 4 years with an average age of 233 days at diagnosis. Only a third (33.3%) of our patients were diagnosed during the neonatal period (before age 7 days). Nine (75%) of our patients presented to our center after age 30 days. No patients were diagnosed between 7-30 days of life (Table 1). Nine patients were males. There were two deaths (mortality = 16.6%).

Prior to admission, most of our patients had been presented to other medical centers with initial complaints of respiratory symptoms, including shortness of breath and cough (Table 2). Six (50% of our patients were diagnosed with pneumonia and as such were treated first with antibiotics before any imaging was performed. Three (25%) of the 12 patients were diagnosed with neonatal sepsis on presentation. One child was diagnosed with congenital pulmonary airway malformation (CPAM). In total, 10 (83.3%) of our patients were initially misdiagnosed. Only one patient had an initial diagnosis of congenital diaphragmatic hernia within the first 10 hours of life. Four (33.3%) patients were found to have associated congenital abnormalities, including Down syndrome (DS) (n=2), inguinal hernia (n=1), Sensorineural hearing loss (SNHL) (n=1). One patient had hypospadias and bilateral undescended testes (Table 3). Eight patients presented with chest X-Rays done in other health facilities.

Two children underwent subsequent chest CT before referral to our hospital. Average age at surgery was 245 days (range 6 days to 4 years). Right sided hernias of all types made up 50% of our cases (n= 6); 16% (n=2) were right sided Morgagni hernias. The most common hernia type was left Bodchalek diagnosed in 5 (41.7%) of cases. There was one left sided diaphragm eventration in the series.

Table 1. Patient Demographics

Case no	Sex	Age at Diagnosis	Age at Surgery
1	M	4 d	9 d
2	F	7.5 m	8 m
3	M	10 h	21 d
4	M	18 d	18 m
5	M	18 m	18 m
6	M	50 d	3 m11 d
7	F	8 m	8 m
8	M	3 d	6 d
9	M	4 y	4 y
10	M	7m	8 m
11	M	38 d	65 d
12	F	4 d	10 d

Key: d = days. h = hours. m = months. y = years

Table 2. Signs, Symptoms, Initial Diagnosis and Modality of diagnosis

Case no	Symptoms	Signs	Initial Dx	Modality
1	Respiratory distress at birth	Diminished breath sounds on L	Perinatal asphyxia (PNA)	Physical exam
2	Cough, SOB, emesis following feeds, diaphoresis	Scaphoid abdomen, nasal flaring, subcostal retraction, decreased air entry	congenital pulmonary airway malformation (CPAM)	Chest CT
3	Respiratory distress	Scaphoid abdomen	CDH	CXR
4	Respiratory distress, difficulty feeding	Dextrocardia	Sepsis	Physical exam
5	Tachypnea, FTT	Diminished breath sounds	PNA, Pneumothorax	CXR
6	Cough, tachypnea, difficulty feeding, projectile vomiting	Decreased air entry bilaterally	Sepsis	Physical exam
7	SOB, fever, cough	absent air entry on left	PNA	Physical exam
8	Respiratory distress	Bowel sounds in left chest	Preterm/Syndromic	Physical exam
9	Cough, weight loss x 3 weeks	Scaphoid abdomen, failure to insert chest tube due to omentum in chest	PNA	CXR
10	Snoring, difficulty breast feeding	Depressed nasal bridge, low set ears, pectus cavatum	PNA	Physical exam
11	Cough, SOB	Bowel sounds in right chest	PNA	Physical exam
12	SOB, fever	Decreased breath sounds	Sepsis	Physical exam

Three (25%) patients were noted to have a portion of the liver herniated into the chest. One patient had associated segment IV liver hypoplasia (Table 4). All hernias were repaired primarily. Mesh patch was used in none of the cases. All repairs were performed through a subcostal abdominal incision. In the immediate post-operative period, two patients required cardiopulmonary resuscitation. Return of spontaneous circulation was achieved in one patient. One patient suffered from a small bowel obstruction, which was managed conservatively, two months post operatively.

One patient developed incisional hernia. One patient was returned to the operating room for a jejuno-jejunal intussusception requiring exploration and manual reduction (Table 4). Average length of stay in the hospital was 24.5 days (range 12-60 days).

Table 3. Associated Congenital Abnormalities

Case no	Congenital abnormalities
1	Right undescended testicle
2	None
3	None
4	Segment IV liver hypoplasia
5	R inguinal hernia
6	None
7	Mosaic Downs Syndrome (DS), bilateral undescended testicles, dysmorphic feature, hypospadias
8	None
9	DS, Sensorineural hearing loss (SNHL)
10	None
11	None
12	None

Table 4. Intraoperative Findings, Type of Hernia, Outcome/Complications

Case no	Intraoperative findings	Type of hernia	Outcome/ Complications
1	Spleen, small bowel in left chest	L Bochdalek	Superficial Surgical Site infection
2	Right lobe of liver in right chest	R Bochdalek	No complications
3	Stomach, transverse colon, spleen in left chest	L Diaphragm eventration	No complications
4	Hernia sac containing spleen, stomach, transverse colon in left chest, splenic capsule bleeding on reduction	L Bochdalek	Jejuno-jejunal intussusception requiring manual reduction
5	Parasternal diaphragmatic defect with ileum and large bowel	R Morgagni	No complications
6	Right lobe of liver, gallbladder, bowel in right chest	R Bochdalek	Prolonged intubation, failed extubation, VAP, cardiac arrest with ROSC after CPR, Kwashkior, Death
7	Large bowel, small intestine in left chest	L Bochdalek	Death
8	Small bowel, colon and spleen in left chest	L Bochdalek	Incisional hernia
9	Large hernia from ribcage to central chest, small bowel, transverse colon, stomach, spleen and left lobe of liver in left chest	L Bochdalek	No complications
10	Anterior diaphragmatic defect of 6x5cm with transverse colon and omentum in chest	R morgagni	No complications
11	Ascending colon and right lobe of liver in chest with hernia sac intact	R Bochdalek	No complications
12	Small bowel, large bowel, stomach and spleen in right chest	R Bochdalek	Partial SBO at 3 months post-op, managed conservatively

Two of our patients died, making a mortality rate of 16.6%. The first death was in a patient who was born to a polyhydramniotic mother. He first presented at age 6 days with tachypnea, difficulty feeding and projectile vomiting. On physical exam, he was noted to have decreased air entry bilaterally. He was initially diagnosed

with neonatal sepsis. He was diagnosed with CDH at age 50 days, after presenting to our center on the 45th day of the life, at which point he was noted to be severely malnourished due to difficulty feeding. He underwent surgery at age of 3 months and 11 days. He was noted to have a right inguinal hernia at birth. Intraoperatively, he was found to have a right Bochdalek hernia, which was repaired primarily through a subcostal abdominal incision. After surgery, he had a prolonged hospital course complicated by failed extubation, ventilator associated pneumonia, cardiac arrest with return of spontaneous circulation after CPR and Kwashiorkor, ultimately dying two months after surgery.

The second death was in a patient who presented at 8 months with shortness of breath, fever, and cough. She presented to an outside facility with similar complaints at age 3 months and was diagnosed with pneumonia. On physical exam, she was found to have absent air entry over the left chest, and was found to have a Left Bochdalek hernia intraoperatively, which was repaired primarily via subcostal abdominal incision. She died two days after surgery. She was never extubated.

Discussion

The literature on CDH out of sub-Saharan Africa is made up largely of single case reports and relatively small case series. Many institutions in the region see an average of one CDH patients per year because of lack of antenatal diagnosis and high mortality in the neonatal period and possible misdiagnosis³. The mortality rate among late presenters in the region remains unknown due to low case numbers. This series of 13 patients contributes the experience of one of the largest tertiary referral centers in sub-Saharan Africa and one of the few with a several full time pediatric surgeons.

CDH tends to have a slight male predominance with a ratio of 1:0.69⁴. Our series was consistent with this trend as 9/12 of our patients were male. Reported rates of other major associated congenital abnormalities in CDH patients vary greatly, but may be as high as 47%⁵. Five (41.7%) of our patients were noted to have associated congenital abnormalities. Two patients (16.7%) in our series were noted to have Down syndrome (DS), which is consistent with reports of trisomy 21 in five to sixteen percent of patients with CDH⁶. One of the patients with DS was noted to have Sensorineural hearing loss (SNHL) at birth. Though SNHL has a known association with CDH and is often diagnosed in the toddler years⁷, audiograms are not routinely performed at our institution

Survival rates among CDH patients vary widely from institution to institution, but 80% is the most often quoted number for patients who are cared for at a tertiary referral center⁸, which is consistent with our overall survival rate of 83.3%, which is most likely slightly higher than the world average as most of our patients are late presenters. The clear majority of the literature comes from institutions with access to Extra Corporal Membrane Oxygenation (ECMO), which is not available in our setting. Patients who present after 30 days are considered late presenters, and are expected to have an excellent prognosis of close to 100% survival once they are properly diagnosed⁹. In our series, two late presenters died.

Historically, most CDH patients died before surgery, either in utero or in the immediate postpartum period. These uncounted cases are considered the “hidden mortality rate” of CDH. Despite worldwide advances in neonatal and antenatal care, the hidden mortality remains constant at approximately 45%¹⁰. We can only assume that the hidden mortality rate in Ethiopia to be much higher, as average age of diagnosis in our series was 232.7 days. Those who present to our center have largely already declared themselves survivors. The majority (66.7%) of patients in our series come from regains outside of Addis Ababa, including Oromiya (n=3), Amhar (n=2), Arada (n=2), and Dessie (n=1) and are referred to TASH after evaluation and initial treatment in their home regions. In our series, 83.3% of patients were initially misdiagnosed, which worsens the problem of hidden mortality as appropriate care is delayed.

CDH diagnostic index in the antenatal period has markedly increased, so that in many high resource countries, most cases are diagnosed during prenatal ultrasound screening⁽¹¹⁾. None of our patients were diagnosed prenatally. Only 25% (n=3) of our patients presented in the neonatal period, defined as prior to 7

days of life. Both patients who died were well outside of the neonatal period on presentation (50 days and 8 months).

In our group, one patient was preterm at delivery. His mother was diagnosed with preeclampsia. The patient was transferred to our neonatal center in the immediate postnatal period for respiratory distress and congenital abnormalities suggestive of Down syndrome, which were recognized on physical exam. CDH was diagnosed at third day of life, and the patient underwent surgery at day 6 of life. He survived with a complication of incisional hernia.

We found no association between age at diagnosis and length of stay or outcome. There was no association between type of hernia and complication rate or length of stay. Our one preterm patient survived, even though patients born preterm have lower chance of survival than full term infants ⁽¹²⁾. We can only assume that most preterm infants with CDH born outside of our hospital never make it to our center, thereby increasing the hidden mortality rate further.

The practice in many neonatal centers is to intubate all patients with CDH on delivery¹³. Because our patients are not diagnosed prenatally, we do not intubate on delivery. In addition, though neonatal care is available throughout Ethiopia, few centers have access to mechanical ventilation. Even at TASH, there are a limited number of neonatal mechanical ventilators, and it is not a routine practice to intubate children on diagnosis. Inhaled nitrous oxide is used routinely along with noninvasive respiratory support as necessary. Though the practice at many institutions is to perform patch repair for certain diaphragmatic defects, at our institution all such hernias are repaired primarily, as evidenced in our series. Most of our patients would not have qualified for patch repair, again suggesting that those patients who make it to our center may have less severe disease.

The major limitation of our series is the relatively small number of patients, which is to be expected given the rare nature of this condition. In addition, our analysis is dependent on handwritten charts as TASH has not established an electronic medical record yet. 20% (3/15) of the charts were unable to be located and were therefore not included in our series. How those patients may have affected our analysis remains unknown.

Conclusion

CDH remains a challenging problem in resource poor countries. Without prenatal diagnosis, the early diagnosis of most vulnerable patients remains dependent on general practitioners and other primary care physicians. We must better equip our doctors and ultrasonographers to make diagnose CDH and other congenital abnormalities prenatally. Health care providers should be better trained to diagnose CDH early and refer patients for appropriate care. Ultimately, said care should be available in centers outside of Addis Ababa, which means increasing pediatric surgical capabilities in other parts of the country. Screening for the CDH should be a routine investigation in all pregnant mothers. In the absence of prenatal diagnosis, CDH should be one of the differential diagnoses in neonates and young children presenting with respiratory distress. Far too many of our patients (50%) were initially diagnosed with pneumonia. Once the diagnosis of CDH has been made or is suspected, patients should immediately be transferred to a tertiary care center for prompt medical management and surgical repair.

Though it has fallen in the past two decades, Ethiopia's fertility rate remains high at 4.6. The infant mortality rate has also remained high at 47 deaths per 1000 live births ¹⁴. We believe that if routine antenatal care and ultrasound screening were performed, we would find that CDH would account for a good number of still births and perinatal deaths. Though religious and social barriers are a hindrance to autopsy in Ethiopia, we do recommend the practice be encouraged, as investigation into cause of death would contribute greatly to our understanding of the epidemiology of CDH and other congenital anomalies.

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