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Single-staged surgical approach in congenital diaphragmatic hernia associated with esophageal atresia

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ABSTRACT

Background: The coexistence of congenital diaphragmatic hernia (CDH) with esophageal atresia (EA) has only been reported occasionally in literature. Series of patients from a single institution with comparison of different postnatal therapeutic approaches have not been reported. We describe our management in this unique cohort of patients and discuss the procedures that can lead to successful outcomes in this association of congenital anomalies.
Methods: The surgical approaches and outcome of six neonates with CDH associated with EA and distal tracheo-esophageal fistula (TEF) are discussed.
Results: Five newborns were treated surgically, while one patient with trisomy 18 only received palliative treatment. In four patients TEF was ligated during laparotomy for CDH repair. Secondary surgery was performed for correction of EA via thoracotomy after 4–6 weeks (primary anastomosis in two patients, Foker's-technique in one patient, one patient deceased prior to secondary surgery). All three surviving patients required fundoplication due to severe gastro-esophageal reflux during the first year of life. Two patients also required dilatation for anastomotic stricture. In one preterm infant correction of both malformations was accomplished during one surgical intervention. The herniated organs were eventrated and temporarily placed into a silastic bag to allow a mediastinal shift to the left. Thus a continuous ventilation of the right lung with minimal compression and sufficient oxygenation was possible during esophageal repair via a right-sided thoracotomy and extrapleural approach. No further surgery was required so far.
Conclusions: Definitive surgical correction in newborns with CDH and EA was so far accomplished with multiple surgical interventions. Ligation of TEF via an abdominal approach with repair of CDH followed by delayed repair of EA is prone to stenosis and gastro-esophageal reflux due to loss of esophageal length. With a new combination of established surgical methods a single-staged correction of both malformations is possible. This new approach might help to preserve sufficient length of esophagus to accomplish primary anastomosis without tension and therefore avoid long-term morbidity and repetitive surgeries.

With an incidence of 1/2500 live births congenital diaphragmatic hernia (CDH) [1] occurs sporadically. The septum transversum separates the thoracic cavity between the 8th and 12th week of gestation. The evagination is primarily composed of pleural membrane and peritoneum, while muscle fibers migrate into this membrane from anterior to posterior. Due to the late closure of this membrane on the left side, left diaphragmatic hernias are more common and are predominantly posterior [2]. As esophageal lengthening occurs at the same time as the growth of the septum transversum the esophagus is prone to developmental disturbances in CDH. Impaired development of the posthepatic mesenchymal plate causes a diaphragmatic defect that is followed by herniation of the liver into the thoracic cavity that further compromises lung development [3,4]. While for isolated CDH a rate of survival till discharge of over 70% is stated [5] the presence of an associated esophageal atresia (EA) with tracheo-esophageal fistula (TEF) is considered to be a fatal combination [6]. The presence of TEF critically limits the possibilities of positive pressure ventilation in neonates with CDH, and the direct connection between the respiratory and the gastrointestinal tract has a decisive role in the urgency of surgical repair [7]. Not much is known about surgical correction of TEF in the context of CDH either, and there is no reliable evidence about treatment strategies. The coexistence of CDH with EA has only been reported occasionally in the literature [6,8] and a quantification of postnatal therapeutic approaches has not been made. We report our management in this unique cohort of patients and discuss the procedures that can lead to successful outcome in this association of congenital anomalies.

1. Case presentations and treatments

CDH is one of the most serious neonatal malformations that are regularly seen in our hospital. The assessment of patients suspicious

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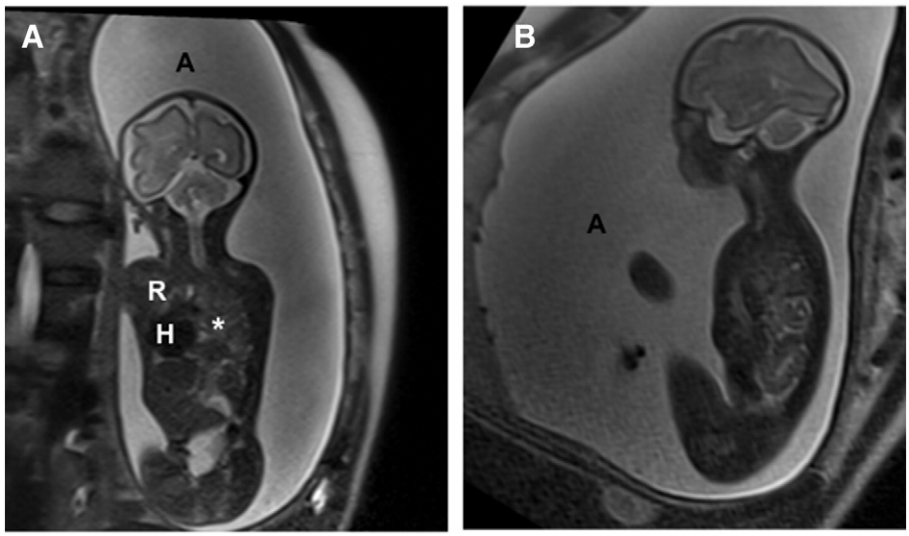

Fig. 1. Intrauterine MRI (26 WOG, patient 6). Coronal and sagittal ultrafast magnetic resonance image of a left-sided congenital diaphragmatic hernia associated with EA at gestational age 26 weeks. Antenatal ultrasound with hydramnios was suspicious for an EA, which could not be confirmed in the MRI presented. **A)** Coronal MRI of a fetus with left diaphragmatic hernia. The herniated intestines are marked with an asterisk (*). **B)** Sagittal MRI of the same fetus with polyhydramnios. The hypopharynx is filled with fluid, the esophagus not definable. L-CDH: left sided diaphragmatic hernia, R: right lung, H: heart, A: amniotic fluid.

for CDH includes prenatal high-resolution ultrasound, fetal MRI scans, echocardiography and genetic testing between 20 and 24 weeks of gestation (WOG). Perinatal management with a well-coordinated multidisciplinary team with extracorporeal membrane oxygenation (ECMO) capability and immediate availability of neonatologists and pediatric surgeons is offered. We favor a planned vaginal delivery around 38 WOG in neonates with a good prognosis (high relative fetal lung volume on antenatal MRI and/or high lung-to-head ratio (LHR >1.6) on ultrasound, 'liver-down' in left-sided CDH) and a Caesarean section in neonates with an LHR <1.6. Determined cut off values for ECMO therapy are an LHR of 1.2 at 34 WOG or fetal lung volume below 25 ml. Cut off values for survival are LHR of 0.9 and fetal lung volume of 9 ml [9]. According to the CDH EURO CONSORTIUM consensus on standardized postnatal management in CDH patients, neonates were immediately intubated and ventilated with low peak pressure (25 cm H_2O or less) to minimize lung injury [10]. A radial or femoral artery catheter is placed for frequent monitoring of blood gases and blood pressure and a central venous catheter for fluids and medication. Surgical repair is delayed until maximal resolution of pulmonary vascular reactivity using strategies like permissive hypercapnia, high frequency oscillation and ECMO [10] if necessary. During the last ten years 520 neonates with CDH with an overall survival rate of 82 % were treated in our hospital. Six (1.15%) neonates had an EA with distal TEF (Type IIIb according to Vogt [11]) in the presence of a unilateral CDH, Bochdalek type hernia. Fetoscopic occlusion of the trachea is not performed in our hospital but cooperation exists and patients are transferred to our hospital for delivery and possible postnatal ECMO. One of the CDH and EA patients received fetoscopic treatment due to rather unfavorable antenatal LHR values and undetected EA. Since 2008 71 neonates with CDH underwent thoracoscopic repair but minimal invasive surgery was not an option in these six neonates.

1.1. Magnetic resonance imaging (MRI)

Fetal MRI in our hospital is performed using a 1.5-T super conducting MRI system (Magnetom Sonata or Avanto, Siemens Healthcare) with a six-element phased-array surface coil. The total fetal body is covered by a HASTE T2-weighted sequence with a 5 mm slice thickness without a gap between 24 and 32 WOG. Imaging is performed in the coronal and sagittal planes relative to the fetus. The examination lasts approximately 15 min. There is no need for fetal sedation. Three fetal patients had an MRI that demonstrated a CDH, while the associated EA with TEF was not detected, Fig. 1. In two patients hydramnios was aspirated that was suspicious for an EA. The EA with TEF was confirmed postnatal, Fig. 2.

1.2. Surgical approach

Five patients were treated surgically, while one neonate with Edwards Syndrome (patient 1) only received palliative treatment. For detailed summary and risk status of all patients see Table 1.

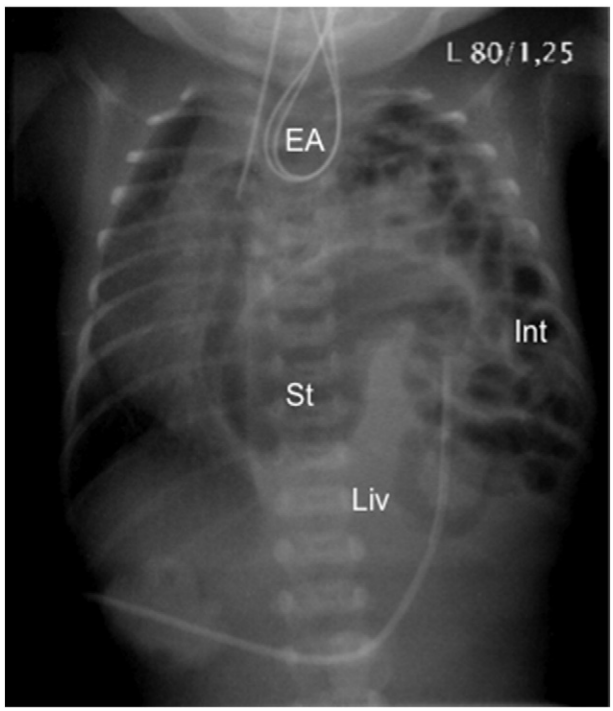

Fig. 2. Postnatal x-ray of the newborn with single-staged EA and CDH repair (patient 6). Postnatal x-ray of the chest within the 1st hour after delivery demonstrates left-sided congenital diaphragmatic hernia (herniated stomach and intestine), esophageal atresia (the nasogastric tube coiled at the level of the T2 vertebra), and conclusively distal tracheoesophageal fistula. The severe mediastinal shift to the right is clearly seen. EA: esophageal atresia, Int: intestines, L: liver, St: stomach.