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Low early and late recurrence-rate after open implantation of a cone-shaped patch for neonatal repair of congenital diaphragmatic hernia – a prospective cohort-study at a single referral centre with a follow-up time of two to ten years

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Congenital diaphragmatic hernia is a rare defect with an incidence of 1:3000 in neonates and patients present complex hemodynamic, respiratory and reconstructive challenges. Nowadays it is still a life-threatening condition and survivors may suffer from long-term-morbidity that also influences quality of life. In large defects a substitute for the diaphragm is required, while different techniques have been described and different patch materials have been used. Recurrence is one of the most severe surgical complications after repair of congenital diaphragmatic hernia with an increased risk of intestinal injury at secondary surgery. In literature incidence varies depending on operative technique, patch material and patient selection between 4% and more than 50%. But studies are difficult to compare, because operative techniques and patch material differ, most are retrospective and do not offer a structured long-term-follow-up to all surviving patients with congenital diaphragmatic hernia. Previously reported recurrence rates therefore certainly have been underestimated. Over the years still high recurrence rates are reported in recent literature after patch repair. This is the largest prospective long-term-study that has been reported from a single institution with a standardized surgical treatment algorithm and a structured long-term-follow-up offered to all surviving patients with a participation-rate of more than 90%. The purpose of this prospective study was to evaluate the impact of surgical techniques on the rate of recurrence in open surgery. Data was collected over a time period of 12 years with a minimal follow-up time of two years to detect incidence and time of recurrence depending on different operative procedures. Between 2003 and 2007 open surgery was performed, since 2008 minimally invasive techniques were also applied. 508 neonates with congenital diaphragmatic hernia were treated at our institution between 2003 and 2012, of whom 200 received extracorporeal membrane oxygenation therapy (ECMO-therapy). 442 patients (87%) underwent surgical closure of the diaphragm, in 47 minimally invasive surgery was applied (10.6%). In open surgery a cone-shaped patch was used in 301 neonates to achieve closure of the diaphragm (76.2%). Overall 410 patients survived (80.7%) and the majority was reassessed in our longitudinal follow-up program also comprising regular radiologic imaging (90.2%).

Early recurrence until discharge was observed in only three of 410 surviving patients (0.7%). In 28 of 326 surviving and reassessed children late diaphragmatic complications occurred after open surgery (8.6%). In six patients multiple recurrences were detected (1.8%). Recurrence at the original localization of the diaphragmatic defect ('true' recurrence) was detected in 24 cases (7.4%), an isolated hiatal hernia in eight (2.5%) and a combination of both in six (1.8%).

Overall 38 diaphragmatic complications were diagnosed (11.7%), 19 in 131 surviving ECMO-patients (14.5%). The rate of diaphragmatic complications after primary repair was 8.8% (6/68 patients) and 12.7% after repair with a cone-shaped patch (32/251 patients). According to defect size higher rates of diaphragmatic complications were detected in larger defects (defect A 0%, defect B 2.8%, defect C 13.1%, defect D 37.5%).

Diaphragmatic complications after repair of congenital diaphragmatic hernia were observed within 12 months after surgery in 21 cases (55.3%), and within 24 months in 32 cases (84.2%).

Long-term-results after diaphragmatic reconstruction highly depend on surgical technique. With the use of a cone-shaped Gore-Tex®-patch long-term overall rate of diaphragmatic complications was 12.7% in open surgery within an observation period of a minimum of two years to a maximum of ten years. As independent variables for diaphragmatic complications of diaphragmatic hernia a left-sided defect and

the need for an abdominal wall patch could be identified as well as a ‘liver-up’-situation in left-sided diaphragmatic hernia.
It is essential to follow these patients in a distinct long-term follow-up program with regular radiologic imaging – since most diaphragmatic complications were detected in patients who showed no or minimal clinical symptoms (97.4%) and because 44.7% of recurrences were diagnosed beyond one year of age. Only a structured follow-up with regular radiologic imaging of all surviving patients with congenital diaphragmatic hernia allows a reliable detection of diaphragmatic complications. If waiting for symptomatic patients, the vast majority will not be diagnosed bearing the risk of intestinal incarceration, bowel gangrene and lethal septicaemia.