

8 Summary

The influence of defect size and surgical technique on the long-term morbidity of children with congenital diaphragmatic hernia

Objective: The aim of this study is to evaluate the effects of diaphragm reconstruction using a synthetic non-absorbable patch from polytetrafluoroethylene (PTFE, Gore-Tex) and primary repair of diaphragm defects on the long-term morbidity in children after surgery of congenital diaphragmatic hernia (CDH), with a primary interest in the influence of these techniques on the occurrence of skeletal deformities and the development of pulmonary functions. One part of this study is to introduce a new objective method for perioperative evaluation of the size of diaphragmatic defects to enable comparison of results among various centres and methods used for diaphragmatic reconstruction. Another part of this study is to evaluate the validity of lung function assessment using four unique methods of Infant Pulmonary Function Tests (IPFT) in children under 13 kg of body weight.

Material and Methods: A study of 69 patients operated on between 1996-2010 for CDH, in which the clinical part of the study was performed between the years 2009-2011. The clinical study evaluated anthropometric examination, pulmonary function testing and a questionnaire study which focused on long-term morbidity and patient quality of life. A prospective study of 47 infants born between 2009 – 2011 was carried out, making up the experimental part of the study which focused on the perioperative measurement of the size of the thorax (chest circumference, sagittal and transverse dimension of the chest) and diaphragm defect size (sagittal and transverse diameter of the defect) to find objective criteria that could indicate the use of a Gore-Tex patch.

Results: Compared with the population norm, CDH children had a significantly lower body weight SDS (mean -0.75, $p < 0.00002$), BMI SDS (mean -0.71, $p < 0.000005$) and lower thoracic index SDS (mean -0.71, $p < 0.00002$). Children after Gore-Tex patch repair (GT) had a significantly higher incidence of pectus excavatum (PE), poor posture (PP) and lower BMI (PE: $p = 0.014$, PP: $p = 0.026$, BMI SDS: $p = 0.009$). Other anthropometric parameters did not significantly differ from children after primary repair

(PR). An increased value of functional residual capacity (FRC) at the age of 1.07 years (median) was found in the GT group compared to PR (165.7 ± 51.9 vs. 120.4 ± 31.2 , $p < 0.02$). Similarly, children with pulmonary hypertension, which were treated in the perinatal period of inhaled nitric oxide (INO), also had significantly higher FRC (183.1 ± 52.6 vs. 117.8 ± 25.7 ml, $p < 0.0005$) at the age of testing, while also exhibiting significantly higher occurrence of pulmonary morbidity. In 34 newborns complete perioperative measurement and categorization of defect size were performed. To determine the relative size of the defect, a defect-diaphragmatic ratio (DDR=defect area: diaphragm area x 100) was calculated. Significant differences in the size of diaphragmatic defect were found between the GT and PR groups with the transverse and sagittal diameters of defect (48.0 ± 5.7 vs. 30.1 ± 5.9 , $p < 0.00061$; 34.0 ± 12.5 vs. 16.0 ± 7.3 , $p < 0.0022$) and DDR index (18.29 ± 4.6 vs. 5.77 ± 3.28 , $p < 0.0005$), respectively. DDR was >15 in all neonates who underwent patch repair in our study.

Conclusion: Reconstruction of large diaphragmatic defects with a synthetic non-absorbable patch (Gore-Tex) remains an acceptable method for the growing child organism. Examination did not show reconstruction with a Gore-Tex patch as being an independent negative factor for lung function development or the formation of skeletal deformities. The calculation of the diaphragmatic defect ratio (DDR) optimizes the indications for use of a Gore-Tex patch. This objective assessment of defect size may be an improvement over the current, more subjective assessment techniques, currently practiced by surgeons, thus providing a standardized criterion which can be of great utility in the field. Infant pulmonary function testing is valid for the assessment of lung development in early childhood. The successful detection of high-risk children allows the early initiation of targeted and effective treatment of asthma. Despite a greater incidence of comorbidities in children after surgery of CDH when compared to the overall population, we need to evaluate that the overall health status of these patients is favorable and that quality of life is maintained.

Keywords: Congenital diaphragmatic hernia, Synthetic non-absorbable patch, Gore-Tex, Anthropometric examination, Infant pulmonary function tests, Skeletal deformity, Lung function, Defect-diaphragmatic ratio, Long-term morbidity, Quality of life.