Journal of Pediatric Surgery (2013) **48**, 935–941

ELSEVIER

Journal of Pediatric Surgery

www.elsevier.com/locate/jpedsurg

Late surgical outcomes among congenital diaphragmatic hernia (CDH) patients: Why long-term follow-up with surgeons is recommended

Tim Jancelewicz, Monping Chiang, Carol Oliveira, Priscilla P. Chiu*

Division of Pediatric Surgery, The Hospital for Sick Children, Toronto, ON, Canada M5G 1X8

Received 21 January 2013; accepted 3 February 2013

Key words:
Congenital diaphragmatic hernia;
CDH;
Recurrence;
Surgical complications;
Long-term outcomes

Abstract

Background: CDH patients experience multi-system morbidity. Despite apparent health, late childhood complications do occur. We reviewed the long-term surgical morbidity of our CDH patients to determine whether protracted clinical surveillance is warranted.

Methods: A single-institution retrospective chart review of all CDH survivors treated from 1999 to 2011 who are followed at our CDH multidisciplinary clinic was performed. Descriptive and statistical analyses were performed to show risk of surgical complications over time.

Results: A total of 187 CDH patients were treated with 160 surviving to discharge (86%). Primary repair was performed in 115 (73%), and 42 (27%) underwent patch repair. CDH recurrence occurred in 23 (15%) at a median time of 0.7 (range 0–8.5) years (65% asymptomatic). Seventy percent of recurrences occurred before 2 years and 17% after 4 years. Bowel obstruction occurred in 12 (8%) at a median time of 0.7 (range 0.2–7.2) years post-repair, and chest deformity occurred in 13 (8%) at a median of 5 (range 1.1–6.8) years. For patch repairs, scoliosis occurred in 4 (10%) patients at a median age of 3 (range 0.6–5) years.

Conclusion: Surgical complications in CDH survivors are common, can occur many years later, and are frequently asymptomatic. Long-term surveillance of CDH patients is recommended for early identification and treatment of complications.

There is increasing evidence that improvements in the survival of patients with severe congenital diaphragmatic hernia (CDH) have been associated with a corresponding increase in the incidence of long-term morbidity [1–3]. Postoperatively, CDH patients face a spectrum of potential complications that require long-term surveillance by clinicians, ideally in a multidisciplinary setting. Even for CDH survivors with minimal pulmonary disease, many centers advocate protocolized out-patient visits with routine imaging so that both medical and surgical concerns may be addressed early, thus minimizing morbidity [4,5].

Unfortunately, despite these recommendations, most CDH patients do not receive structured follow-up and may not be seen by a surgeon routinely [6]. Additionally, it is not clear when or even if such careful follow-up should conclude, as truly long-term outcome patterns are not well described in the literature. Indeed, reports differ on the

* Corresponding author. Tel.: +1416 813 6405.
E-mail address: priscilla.chiu@sickkids.ca (P.P. Chiu).

0022-3468/$ – see front matter © 2013 Elsevier Inc. All rights reserved.
http://dx.doi.org/10.1016/j.jpedsurg.2013.02.005

definition of "long-term outcomes," with some groups reporting 1 or 2 years of follow-up data as "long-term" complication rates [7,8]. Still other groups quote low hernia recurrence rates without complete follow-up of their populations in clinic or without routine surveillance imaging of healthy survivors [9]. Such perspectives carry the implicit suggestion that close follow-up is unnecessary in healthy children with CDH beyond 12–24 months of age. However, a number of studies have shown that surgical complications such as scoliosis, chest wall deformity and bowel obstruction occur years after CDH repair, and are more delayed or gradual in onset than the physiologically significant adverse nutritional, gastrointestinal, and neurodevelopmental conditions experienced by some CDH survivors [10–17]. Additionally, long-term CDH recurrence patterns are not well defined for three reasons: first, because many survivors of severe CDH defects are still relatively young; second, the durability of prosthetic patch repairs during the rapid growth phases of late childhood and adolescence has not been widely reported; and finally, there is a lack of long-term follow-up by surgeons in many CDH centers.

Since 2000, CDH patients have been followed at our center in a multidisciplinary clinic with a follow-up protocol that includes routine surveillance imaging in accordance with published recommendations [1,5,6]. In Canada, access to CDH clinic is not limited by financial or insurance concerns, allowing us to see virtually all our CDH families in follow-up without undo resource burden. The purpose of this study was to describe patterns of long-term surgical morbidity seen in our cohort of CDH survivors, in order to define the long-term surgical burden of disease for CDH patients and to determine the need for long-term surveillance by surgeons along with other specialists.

1. Methods

Permission for this study was obtained from the Research Ethics Board of the Hospital for Sick Children (1000010450).

1.1. Study cohort

A retrospective review was performed of all CDH patients currently followed in multidisciplinary clinic at The Hospital for Sick Children, University of Toronto, with a date of birth in or after January 2000. Patients born after September 2011 were excluded from the analysis to limit bias due to short follow-up time. All patients are followed prospectively in CDH clinic, and undergo a chest x-ray at each visit, as surveillance for CDH recurrence.

Clinical, operative, and outcomes data were tabulated for subgroup comparison. Specifically, the timing and clinical features of all surgical complications were documented including hernia recurrence requiring operative repair, small bowel obstruction (SBO) requiring laparotomy, major chest wall deformity (defect sufficient to plan referral for eventual treatment), and scoliosis as assessed by physical exam and sequential imaging.

1.2. Operative repair

Prior to 2005, patients underwent open repair with primary closure, muscular flap closure using the method of Simpson as previously reported [18,19], or patch closure using polytetrafluoroethylene (PTFE, Gore-Tex® or Gore® Dualmesh®, W. L. Gore and Associates, Flagstaff, AZ) or Surgisis® (SIS, Cook Medical, Bloomington, IN). After 2005, minimally invasive (MIS) repair (usually thoracoscopic) was employed in select patients without pulmonary hypertension, and the Simpson flap was no longer used due to abdominal wall deformity. Currently, patch repair is performed using PTFE at our institution due to published reports and our own observations of higher recurrence rates with bioabsorbable mesh [3].

1.3. Statistical analysis

Group comparison was made using Fisher's exact test for categorical variables, and Student's t test for continuous variables, with a $P<0.05$ considered significant. Since CDH recurrence can occur at any time post-repair, Kaplan–Meier survival analysis was used to assess and visually demonstrate the chronology of CDH recurrence in the CDH cohort.

2. Results

2.1. Study cohort

From January 2000 to September 2011, 187 patients were treated for CDH at our institution, with 160 surviving to discharge (86%). There have been no post-discharge deaths in the cohort. Three patients were excluded due to lack of follow-up, leaving a study cohort of 157 patients. Of these, 115 (73%) underwent primary repair, and 42 (27%) had a patch repair. Clinical characteristics are shown in Table 1, with comparison between patch and primary repair subgroups.

2.2. Recurrence data

A total of 23 patients had a CDH recurrence requiring operation for repair, yielding an overall recurrence rate of 15%. Recurrence rates stratified by repair type are presented in Table 2. The MIS group had a higher recurrence rate (32%) than patients with open repairs (11%); excluding all MIS repairs, the cohort recurrence rate was 4% for primary repairs and 27% for patch repairs.

CDH recurrences were sometimes seen on surveillance x-ray at clinic in asymptomatic or minimally symptomatic patients, leading to elective repair; this was the case in 15