Seminars in Fetal & Neonatal Medicine 19 (2014) 385–391

Contents lists available at ScienceDirect

Seminars in Fetal & Neonatal Medicine

journal homepage: www.elsevier.com/locate/siny

Review

Multidisciplinary long-term follow-up of congenital diaphragmatic hernia: A growing trend

Sarah Tracy [a], Catherine Chen [b,*]

[a] Department of Surgery, Beth Israel Deaconess Medical Center, Boston, MA, USA
[b] Department of Surgery, Boston Children's Hospital, Boston, MA, USA

Keywords:
Congenital diaphragmatic hernia
Multidisciplinary
Long term follow-up

SUMMARY

This literature review discusses the growing trend toward multidisciplinary long term follow-up for congenital diaphragmatic hernia.

1. Introduction: History and rationale for long-term follow-up

Over the past two decades, improvements in ventilation strategies and prenatal diagnosis have led to increased survival rates of infants born with congenital diaphragmatic hernia (CDH) [1,2]. However, survival has come with a cost. Many survivors have significant short-term and long-term morbidity [1,3–12]. Pulmonary sequelae, gastrointestinal disorders, hearing loss, neurodevelopmental deficits, and other abnormalities are frequently reported among survivors, especially during the first three years of life. A consensus seems to have been reached: these are often complex patients who could benefit from long term follow-up in multidisciplinary clinics [13]. Boston Children's Hospital established the first multidisciplinary CDH clinic in 1990 [9], and as Boston and other centers began to report on the "hidden morbidity" of CDH in the 1990s, it became a recognizable trend to establish centralized long term follow-up to best serve CDH patients and their families [1,9,10]. Since the late 1990s, other multidisciplinary CDH clinics have been established in the USA, Canada, Europe, and other major cities around the world. Follow-up protocols vary substantially among existing clinics, as noted by Safavi et al. across Canadian centers [12]. Some pediatric multidisciplinary clinics have begun to accept referrals from pediatric surgeons for CDH patients even though they were originally established in the 1970s and 1980s to follow low birth weight infants. Pediatric surgeons and other providers now recognize that more CDH survivors are "high risk" infants predicted to suffer from developmental delay, failure to thrive, and chronic pulmonary disease.

As more survivors reach school age, adolescence and young adulthood, several centers have continued to follow these patients in order to better understand the functional status and incidence of chronic medical issues in these populations [3,4,11,13–17]. Many centers also recognize the value of protocol use and prospective evaluation: as treatment strategies evolve and new screening strategies are employed, outcomes will likely change. For example, Chiu et al. found that adoption of a new gentle ventilation protocol was associated with significant improvements in survival rates of CDH patients, but also with increased bronchodilator prescriptions and gastrostomy tube placements (surrogates for pulmonary and nutritional morbidity) three years post-CDH repair [1]. Longitudinal reports from high-volume centers with CDH clinics will better enable providers to identify trends and associations, such as possible increasing morbidity in some of today's more severely affected survivors.

The American Academy of Pediatrics (AAP) published recommendations for a multidisciplinary clinic follow-up schedule for CDH patients in 2008 [18]. The report notes that the published schedule is most applicable to "children with extraordinary medical and surgical complications associated with CDH," and that it should be individualized depending on the specific needs of each infant. This still leaves a lot of room for interpretation and allows providers to use their best clinical judgment. There is little evidence-based reporting on the long-term prognostic value of studies often utilized in CDH follow-up clinics, such as neuroimaging [19], developmental screening, neurodevelopmental evaluation, ventilation/perfusion (V/Q) scanning, pulmonary function testing, chest X-rays, echocardiograms, and upper gastrointestinal endoscopy. Clinics often operate as investigational units as well as patient care centers. Many submit clinical outcomes data to the CDH Study Group registry, which was established in 1995 [20]. CDH clinics are beginning to participate in multicenter trials and employ their own testing and screening initiatives to further understand what is happening

* Corresponding author. Address: Department of Surgery, Boston Children's Hospital, 300 Longwood Avenue, Boston, MA 02115, USA. Tel.: +1 617 355 0535; fax: +1 617 730 0298.
E-mail address: catherine.chen@childrens.harvard.edu (C. Chen).

http://dx.doi.org/10.1016/j.siny.2014.09.001
1744-165X/

with their cohorts [12,21,22]. Such multi-institutional studies will allow investigators to amass larger cohorts of patients and assess the impact of contemporary management strategies on clinical outcomes. Multidisciplinary clinics also facilitate the acquisition of data to correlate patient and parent perspectives with objective findings on pulmonary, gastrointestinal, or neurodevelopmental testing [23–25]. Ultimately, assessing patient and parent well-being remains a primary goal [11,26,27].

2. Description of team members and algorithm for long term follow-up

Table 1 lists all known medical centers to date that have established multidisciplinary CDH follow-up clinics and that have published long-term outcomes data in the literature. As noted previously, some hospitals, such as Montreal Children's Hospital, refer CDH patients to larger "high risk" infant follow-up programs, which include pre-term and very low birth weight infants [28]. At some institutions, it is unclear whether all CDH survivors are referred to these follow-up programs, or whether only CDH survivors stratified as "high risk" receive referrals. The University of California at San Francisco offers two separate follow-up pathways for CDH patients based on acuity or risk [2]. They categorize patients as low acuity if they have undergone primary repair and avoided extracorporeal membrane oxygenation (ECMO) therapy, are discharged on nipple feeds and without supplemental oxygen, and are growing and developing appropriately at their first follow-up appointment after hospital discharge. Patients who do not meet these criteria are classified as complex, and are seen more frequently than the low acuity patients, especially during the first year of life [2].

Although follow-up protocols vary by institution, team membership is relatively consistent. A pediatric surgeon, cardiologist, pulmonologist, gastroenterologist, and developmental pediatrician usually make up the core subspecialty team. Physical and occupational therapists, nutritionists, nurses, and pediatric subspecialists are included in nearly every clinic. Additional staff members may include clinical geneticists, neonatologists, psychologists, orthopedic surgeons, and otolaryngologists [5,6,12,36,37,47,55–58]. Several institutions also report that they have a social worker involved in their multidisciplinary clinic. Based on recent studies, there clearly is an impact on family, even years after initial repair, associated with the care and potential long-term morbidity of CDH, and many families could benefit from social work services in the outpatient clinic setting [11,26].

In 2008, the AAP published an algorithm to help guide follow-up of complex CDH patients for clinicians who may not have access to a multidisciplinary clinic [18]. This document contains a time line that outlines recommended studies to evaluate and treat pulmonary morbidity, but also to screen for foregut dysmotility, growth failure, neurocognitive delay, behavioral disorders, hearing loss, late surgical complications and orthopedic deformities. The AAP further recommends that multidisciplinary clinics follow complex CDH patients through age 16 years. Follow-up through adolescence is especially important for patients with prosthetic patch repairs. These patients have been shown to be at higher risk for the development of pectus excavatum, scoliosis, and other musculoskeletal deformities, and patients with these deformities may experience functional limitations during adolescence as they undergo periods of rapid growth [42].

Table 1
Medical centers with established multidisciplinary congenital diaphragmatic hernia follow-up clinics and long-term outcome publications.

Medical center	Year established	No. of long-term outcome publications 1990–2014							
		Total[a]	Cardiopulmonary	Gastrointestinal/ nutrition	Early neurodevelopment 0–5 years	Hearing loss	Late surgical	School-age function and well-being	Impact on family
Boston Children's Hospital, Boston, MA, USA	1990 [9]	13	3 [5,29,30]	2 [6,9]	4 [9,31–33]	3 [9,34,35]	1 [9]	1 [11]	2 [26,27]
Hospital de São João, Porto, Portugal	1997 [36]	1	1 [36]	1 [36]	1 [36]	1 [36]	–	–	–
Erasmus Medical Center – Sophia Children's Hospital, Rotterdam, The Netherlands	1999 [8]	8	2 [8,14]	1 [37]	2 [14,37]	–	–	5 [13,15,16,38,39]	–
Hospital for Sick Children, Toronto, ON, Canada	2000 [10]	6	4 [1,12,23,40]	2 [1,12]	2 [1,12]	1 [12]	3 [1,10,12]	1 [41]	–
UCSF Medical Center, San Francisco, CA, USA	2000 [42]	2	1 [2]	1 [2]	1 [2]	1 [2]	1 [42]	–	–
Children's Hospital of Philadelphia, PA, USA	2004 [43]	4	1 [44]	–	2 [43,46]	1 [45]	–	–	–
Bambino Gesù Children's Research Hospital, Rome, Italy	2004 [47]	2	2 [47,48]	1 [47]	–	1 [48]	1 [47]	–	–
Morgan Stanley Children's Hospital of New York at Columbia University Medical Center, New York, NY, USA	2005 [21]	6	2 [21,49]	1 [50]	1 [22]	–	2 [51,52]	–	–
Mott Children's Hospital, University of Michigan Health System, Ann Arbor, MI, USA	2008 [53]	3	2 [21,53]	1 [53]	1 [22]	1 [53]	_	1 [53]	–
Alder Hey Children's Hospital, Liverpool, UK	Prior to 2013 [54]	1	–	–	–	–	1 [54]	–	–

[a] The total may not equal the sum of the subcategories, as some publications report on multiple outcomes.