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Musculoskeletal deformities following neonatal thoracotomy: long-term follow-up of an esophageal atresia cohort

Sunny Wei [a], Neil Saran [b,c], Sherif Emil [a,c,*]

[a] Division of Pediatric General and Thoracic Surgery, The Montreal Children's Hospital, McGill University Health Centre, Montreal, Quebec, Canada
[b] Division of Pediatric Orthopaedics, The Montreal Children's Hospital, McGill University Health Centre, Montreal, Quebec, Canada
[c] Chest Wall Anomalies Center, Shriners Hospital for Children, Canada, Montreal, Quebec, Canada

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ABSTRACT

Background: Musculoskeletal deformities (MD), including scoliosis and chest wall anomalies, are potential long-term complications of neonatal thoracotomies.
Methods: We studied the incidence of MD in patients who underwent open repair of esophageal atresia between 1997 and 2012, had no other predisposition to MD, and subsequently received longitudinal follow-up in a multidisciplinary esophageal atresia clinic. Detailed chest wall and musculoskeletal exams were performed at each visit. Incident rate and incident rate ratios were used to determine the incidence of deformities. Logistic regression methods were used to test the effect of independent variables including sex, gestational age, muscle division, number of thoracotomies, and operative complications on the occurrence of MD.
Results: The study cohort consisted of 52 patients followed for a median of 8 (range 1–19) years. MD developed in 13 (25%), with an incident rate of 2.92 per 100 child-years. Division of the serratus anterior was associated with a significantly higher probability of developing MD (log-rank $p = .0237$) and was also a strong predictor of the same [OR 8.6 (95% CI 1.8–42.1)] after adjusting for possible confounders.
Conclusions: Musculoskeletal deformities develop in a significant proportion of neonates following thoracotomy. A muscle-sparing technique decreases the incidence of these deformities.
Type of study: Prospective Cohort Study.
Level of evidence: II.

Repair of esophageal atresia with or without trachea-esophageal fistula (EATEF) is one of the most frequent indications for thoracic surgery in neonates. Until recently, a right posterolateral thoracotomy has been the standard surgical approach [1,2]. Since its inception, there has been ample evidence associating neonatal thoracotomy with long-term thoracic musculoskeletal deformities, including scoliosis, chest wall deformities, and scapular anomalies, with a prevalence of up to 50% [2–8]. However, these studies typically reflect older thoracotomy techniques that included muscle division, and tight chest wall closure with potential rib fusion.

Atrophy of the serratus anterior, as the result of its division, has been implicated in many of the musculoskeletal sequels associated with thoracotomies [6,7,9]. Modern neonatal and pediatric thoracotomy techniques, therefore, attempt to preserve the major chest wall muscles, and avoid rib resection and fusion. A compounding factor in this discussion is the rising popularity of thoracoscopic surgery for repair or EATEF in the past 15 years. Decreased post-operative pain and improved cosmetic results have been presented as advantages of these procedures. However, the main benefit has been the assumption that these procedures decrease or eliminate musculoskeletal deformities [10,11]. This assumption has been made after comparison of thoracoscopic results with studies of open surgery published in the 1980 and 1990's [10,11].

Despite the evolution of neonatal thoracotomies towards muscle-sparing techniques, there have been few studies evaluating whether the risks of long-term musculoskeletal morbidities associated with thoracotomies are still valid in the modern era. As such, the purpose of our

* Corresponding author at: Division of Pediatric General and Thoracic Surgery, The Montreal Children's Hospital, 1001 Decarie Boulevard, Room B04.2028, Montreal, Quebec H4A 3J1, Canada. Tel.: +1 514 412 4497; fax: +1 514 412 4289.
E-mail address: Sherif.Emil@McGill.ca (S. Emil).

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study was to analyze the incidence, type, and severity of musculoskeletal deformities in children who have undergone a thoracotomy for the correction of EATEF in the last 20 years. Longitudinal long-term follow up in a multidisciplinary EATEF clinic, and the existence of a free-standing children's hospital partnering with a Shriner's orthopedic hospital, provided an ideal setting to study these outcomes.

1. Methods

1.1. Study population

The study population consisted of children who had undergone correction of EATEF during the 15-year period between January 1, 1997 and January 1, 2012, and who survived for a minimum of 3 years following the procedure. During that period, all repairs of EATEF were performed through a right posterolateral thoracotomy unless a right-sided descending aorta was seen on pre-operative echocardiogram. Patients with concomitant spinal or chest wall anomalies present at the time of surgery, or who had subsequent diagnoses or procedures associated with the risk of spinal or chest wall anomalies (e.g. cerebral palsy, congenital diaphragmatic hernia, butterfly vertebra, sternotomy for repair of congenital heart defect) were excluded.

1.2. Patient follow-up

At our institution, most patients with esophageal atresia are followed in a multi-disciplinary clinic staffed by pediatric gastroenterologists, pediatric surgeons, pediatric respirologists, pediatricians, dieticians, advance practice nurses, and a nutrition psychologist [12]. The clinic was started in 2006, but all patients with esophageal atresia who were still younger than 18 years were enrolled. At each follow-up visit, a musculoskeletal exam is performed with specific screening and documentation of scoliosis, scapular anomalies (winged scapula), chest wall asymmetry, and chest wall anomalies (pectus carinatum or pectus excavatum). The diagnoses of musculoskeletal deformities were made clinically. If there was significant clinical suspicion, accompanying imaging (i.e. scoliosis series) was included as well. A unique feature of our clinical setting is a freestanding children's hospital and a partner Shriners orthopedic hospital that serves as a regional referral centre for musculoskeletal deformities. The same teams of orthopedic surgeons, as well as surgical and medical subspecialists, staff both hospitals. Patients with scoliosis or chest wall anomalies following thoracotomy are seen at the Shriners Hospital by orthopedic surgery only in the case of scoliosis, or in a multidisciplinary chest wall anomaly center in the case of chest wall anomalies.

1.3. Patient data

For each patient, three potential sources of clinical data were retrospectively reviewed: the Montreal Children's Hospital inpatient chart, the Montreal Children's Hospital outpatient chart including the longitudinally collected EATEF clinic notes, and the Shriners Hospital chart, if one existed. Data collected included patient characteristics, operative details documented in the operative procedure note and operative report (specifically detailing whether the chest wall muscles were divided, retracted, or dis-inserted during the operative approach), post-operative outcomes, and long-term follow-up, as it pertains to the development of spinal and chest wall anomalies. The division or preservation of the latissmus dorsi was not clearly documented in the majority of operative reports and could not be used in the analyses, whereas the division or preservation of the serratus anterior was clearly documented in the majority of reports. Data specific to the long-term follow-up and management of musculoskeletal issues in patients who were assessed at Shriners Hospital were extracted as well.

1.4. Statistical analyses

The Biostatistics Service of the Centre for Innovative Medicine, McGill University Health Centre Research Institute, performed all statistical analyses. The variables analyzed as potential determinants of musculoskeletal deformities included sex, gestational age, muscle division during thoracotomy, number of thoracotomies, intra-operative complications, use of a chest tube, duration of chest tube drainage, and hospital stay. Chi-squared tests and rank-sum tests were used to investigate an association between the development of musculoskeletal deformity and the categorical or continuous variables listed above. Incidence rates (IR), and incidence rate ratios (IRR) were used to estimate the incidence of deformities. The incidence rate was calculated in child-years in order to better account for the fact that the amount of observation time differed between patients in our cohort. It is understood that, in children, the incidence rate of musculoskeletal deformities may increase over time, given the developmental processes in place. As such, we also presented incidence data in a plot of cumulative incidence in order to better represent the changes in incidence rate over time. The incidence rate ratio was used to demonstrate the fold-increase in the incidence of musculoskeletal deformities with division of the serratus anterior. Logistic regression methods were used to examine the potential effect of gestational age, sex, operative complications, and serratus division on the occurrence of musculoskeletal deformities. The log-rank test was used to compare the development of musculoskeletal deformities over time in patients in which the serratus anterior was spared and divided.

1.5. Study approvals

The study was approved by the Research Ethics Board of the McGill University Health Centre (14–530 PED) and the McGill University Faculty of Medicine Institutional Review Board (A07-M41-15B), the approving body for Shriners Hospital studies.

2. Results

2.1. Patient cohort

The patient cohort studied is shown in Fig. 1. A search through medical records by diagnosis and procedure codes, supplemented by the EATEF clinic database, produced 84 patients who were diagnosed with EATEF at the Montreal Children's Hospital during the period of interest. The EATEF clinic database produced an additional 5 patients who had repairs done at other centers, for a total of 89 patients potentially eligible for the study. Nineteen patients were excluded, as shown in Fig. 1, because of concomitant anomalies or procedures predisposing to musculoskeletal deformities. Eight additional were excluded due to a failure to meet the 3-year survival minimum. Of these 8 patients, 1 died intra-operatively secondary to misplacement of an endotracheal tube, and four died during the postoperative period (one from sepsis, one due to esophageal leak with tension pneumothorax, one following complex cardiac surgery, and one from an unidentified cause). Care was withdrawn from two syndromic patients with severe comorbidities upon the parents' request. One patient died of uncontrolled hemorrhage during the second year of life when an esophageal stent eroded into a previously unrecognized aberrant right subclavian artery. Of 62 patients who met inclusion and exclusion criteria, 59 (95%) were operated at our institution by one of seven pediatric surgeons and 3 operated elsewhere. Ten patients did not have adequate follow-up musculoskeletal exams and were excluded from further analysis. Of the 52 patients who constituted the final study cohort, 45 (87%) were followed in the EATEF clinic and 7 (13%) in other clinics. All had good longitudinal documentation of the musculoskeletal exam. The follow-up period was a median of 8 years (range 1–19 years). Fifty patients (96%) were followed for more than 3 years after thoracotomy and 15 (29%) were followed after age 12 years.