Musculoskeletal deformities following repair of large congenital diaphragmatic hernias

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ABSTRACT

Purpose: Large congenital diaphragmatic hernias (CDH) can be repaired with either a muscle flap or prosthetic patch. The purpose of this study was to assess the frequency and severity of scoliosis, chest wall, and abdominal wall deformities following these repairs.
Methods: Neonates who underwent CDH repair (1989–2012) were retrospectively reviewed. We then validated our retrospective review by comparing results of a focused radiologic evaluation and clinical examination of patients with large defects seen in prospective follow-up clinic. Tests for association were made using Fisher's exact test.
Results: 236 patients survived at least 1 year. Of these patients, 30 had a muscle flap, and 13 had a patch repair. Retrospectively, we identified pectus in 9% of primary repairs, 47% of flap repairs, and 54% of patch repairs. We identified scoliosis in 7% of primary repairs, 13% of flap repairs, and 15% of patch repairs. Prospectively, 75% of flap patients and 67% of patch patients had pectus and 13% of flap patients and 33% of patch patients had scoliosis. There was no significant difference between flap and patch patients.
Conclusions: Scoliosis and pectus deformity were common in children with large CDH. The operative technique did not appear to affect the incidence of subsequent skeletal deformity.

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Large congenital diaphragmatic hernias (CDHs) require repair with either a patch or an autologous tissue transfer. Repair with a prosthetic patch is the technique used by most surgeons [1]. Multiple studies have shown that patients who undergo CDH repair with patch or muscle flap have an increased incidence of chest wall deformities, with a scoliosis rate that ranges from 4% to 50% and a pectus deformity rate that ranges from 14% to 80% [2–8]. Repairing large defects with autologous tissue in the form of a split abdominal wall muscle flap is the treatment of choice at our institution, and we have previously reported a low recurrence rate associated with this repair type (4.3%) [9]. The question arises as to whether this type of repair increases abdominal wall weakness or hernia and chest wall deformities. We reviewed our series with a focus on these issues.

1. Methods

1.1. Study population

After obtaining approval from the Institutional Review Board, a retrospective review of all children with CDH repair at our regional tertiary care children's hospital from 1989 to 2012 was performed.

The patients were categorized by the technique of their repair. Repair types included primary repair, split abdominal wall muscle flap and synthetic patch. Paper and electronic medical records were reviewed to obtain demographic data, and diagnosis of skeletal deformities as well as any treatment for the skeletal deformities. Electronic charts were searched for the key words "pectus" and "scoliosis," and those specific notes were reviewed. These diagnoses were made by a variety of physicians including radiologists, orthopedists and primary care physicians and were not always confirmed by a focused follow-up visit by a pediatric surgeon.

In order to check the validity of our retrospective review, patients with large defects were seen prospectively for focused follow-up and a single pediatric radiologist (G.H.) reviewed the most current chest radiograph to evaluate for scoliosis greater than 10 degrees. The results of follow-up were correlated with our retrospective review.

1.2. Operative technique

The choice of surgical technique for repair of the large CDH was according to surgeon preference. In both techniques, as much of the native diaphragm as possible is closed primarily using pledgeted sutures. In patch repairs, the patch is tailored with adequate redundancy to allow tension free growth. Patches are fixed to the margins of the diaphragm if possible or to the pericostal tissue. Split abdominal wall muscle flaps are done using the technique that we

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Table 1
Results of retrospective chart review.

	n	Pectus Deformity	Pectus Deformity RR (95% CI, p)	Scoliosis	Scoliosis RR (95 %CI, p)
Primary Repair	193	17 (8.8%)	Ref.	13 (6.7%)	Ref.
Large Defect	43	21 (49%)	5.5 (3.2–9.6, p < 0.001)	6 (14%)	2.1 (0.83–5.1, p = 0.07)
Muscle Flap	30	14 (47%)	5.3 (2.9–9.6, p < 0.001)	4 (13%)	2.0 (0.69–5.7, p = 0.26)
Patch	13	7 (54%)	6.1 (3.1–12.0, p < 0.001)	2 (15%)	2.3 (0.58–9.1, p = 0.24)

have previously described using the transversalis and internal oblique muscles [10].

1.3. Follow-up

Primary Children's Hospital is the only free standing children's hospital serving the state of Utah and a significant portion of surrounding states in the Intermountain West. Within our catchment area, children receive all of their tertiary care at our facility. In addition to this, Primary Children's Hospital is part of Intermountain Healthcare that has a system wide electronic medical record. Therefore we are able to access ongoing records even in children who have not followed up in the pediatric surgery clinic. In response to recent guidelines [11], all children with large defects have been contacted to return for long-term follow-up.

1.4. Statistical analysis

Tests for association were made using Fisher's exact test, with p-values adjusted for multiple comparisons. All analyses were performed using SAS 9.2 software.

2. Results

Two hundred and seventy-nine infants had CDH repair during the study period and 236 patients survived to at least 1 year of life or were alive and less than 1 year old at the time of review. Of these survivors, 193 underwent primary repair, 30 had a split abdominal wall muscle flap repair and 13 had a patch repair. At the time of chart review the median age of patients who underwent primary repair was 11 years (range 0.5–24.2 years), flap repair was 7.1 years (range 0.8–22.3 years), and patch repair was 4.5 years (0.9–23.8 years).

Results of the retrospective review are summarized in Table 1. Children who had defects that could be closed primarily were significantly less likely to develop chest wall deformities than those with large defects that required either muscle flap or patch repairs (RR 5.5, 3.2–9.6). There was a trend towards an increased risk of scoliosis in children with large defects but this did not reach significance (RR 2.1, 0.83–5.1). The prevalence of chest wall deformity and scoliosis was not different between the muscle flap and patch groups. No child who underwent repair of a large CDH has required operative intervention for a musculoskeletal deformity at this time. One child who underwent primary CDH repair has had a Nuss Procedure for pectus excavatum, and 3 children, all of whom had primary CDH repair, have undergone operative intervention for

scoliosis. Three additional children have undergone bracing for scoliosis, 2 after primary CDH repair and 1 flap patient.

Table 2 compares the results of the chart review with our focused prospective clinic and focused radiological review. Eleven patients with large defects repaired by either split abdominal wall muscle flap or prosthetic patch participated in a long-term follow-up clinic that included a chest x-ray at a median age of 7.8 years (range 3.3–16.3 years). Most chest wall deformities were identified through the chart review with two more being identified prospectively. The chart review sensitivity was therefore 75% for chest wall deformity. Of note, the prospectively identified deformities were characterized as mild by the surgeon and not expected to require repair. The focused clinic visit and chart review identified identical patients with scoliosis. In contrast, review of current chest radiographs looking for scoliosis defined as a curvature of 10 degrees or more identified 2 additional patients. Both the chart review and clinical examination had a sensitivity of 50% for this degree of scoliosis. A single radiologist also reviewed the most recent available chest radiographs (26) for patients with large defects at a median radiologic follow-up of 1.5 years. This review identified scoliosis in 30% of the flap group and 33% of the patch group, similar to detailed review of x-rays from follow-up clinic.

The patients who presented for focused, prospective follow-up were also evaluated for abdominal wall hernia. Although protrusion of the abdominal wall is always seen following muscle flap repair in the short term, this defect improves over time and currently only 2 (25%) patients have a bulge. No patient has required operative intervention for a ventral hernia.

3. Discussion

Congenital diaphragmatic hernia is a severe developmental defect that is characterized by not only a defect in the diaphragm but also by ipsilateral and contralateral pulmonary hypoplasia. Given the close embryologic relationship between the lungs, thoracic cage and diaphragm, it is reasonable to expect chest wall and thoracic spine deformities in patients with CDH [5,12]. Our study provides an estimate of this risk. As hypothesized, more severe diaphragmatic defects are associated with a significantly greater risk of pectus deformities, and a trend toward increased scoliosis. This risk seems to be independent of the type of repair employed, supporting the notion that this may arise from the underlying developmental abnormality or tension inherent in any type of repair performed on a large defect.

It has been postulated that the tension after repair of large CDH defects may lead to scoliosis [5]. It has also been suggested that patch repairs actually decrease tension compared to primary repair, thus

Table 2
Accuracy of Retrospective Chart Review Compared to Focused Prospective Clinic Visit.

	Pectus Deformity		Scoliosis		
	Focused Prospective Clinic Visit	Chart Review	Focused Prospective Clinic Visit	Chart Review	Focused Radiology Review
All (n = 11)	8 (73%)	6 (55%)	2 (18%)	2 (18%)	4 (36%)
Muscle Flap (n = 8)	6 (75%)	5 (63%)	1 (13%)	1 (13%)	3 (38%)
Patch (n = 3)	2 (67%)	1 (33%)	1 (33%)	1 (33%)	1 (33%)

implying that these repairs may be beneficial in preventing abnormal musculoskeletal development [13]. In patients with a large CDH, the lungs are similar to those of premature infants demonstrating decreased surfactant, poor compliance and immature morphology [14,15]. To overcome poor compliance a greater negative intrathoracic pressure is needed which could theoretically cause inward pull of the chest at its compliant anterior cartilaginous wall [5]. These musculoskeletal deformities can be thought of as primarily associated with CDH or as secondary deformities resulting from the patient's physiology and the tension of the repair. Though many theories for the increased incidence of pectus deformity and scoliosis have been proposed, a clear explanation is yet accepted.

Scoliosis, defined as a lateral curvature of the spine greater than 10° accompanied by vertebral rotation, is present in 2% to 4% of adolescents in the general population [16]. The prevalence of a pectus deformity in adolescents is approximately 1% [2,17]. In addition, these two chest wall deformities are often seen in association, out of a group of 248 children without CDH but with pectus excavatum, the prevalence of scoliosis was 23% [18], much higher than what would be expected in the general population.

Several studies have described an increase in chest wall deformities following CDH repair [3–8,19–24]. In children with large defects, scoliosis has been described in between 4% and 50% of children, and pectus deformity in between 14% and 80% of those undergoing patch or flap repair. Nasr et al. compared chest wall deformities in patients repaired by flap and patch and found no statistical difference across treatment types. In their series, 16% who underwent flap repair developed an abdominal wall defect at the flap donor site but none required surgery. They concluded that both flap and patch repair provide similar long-term results [3]. Our study demonstrated pectus deformity in 49% of children requiring patch or flap repair, and scoliosis in 14%. These may be underestimates based on the results of our focused prospective clinic visits and radiologic evaluation. While we were not able to demonstrate a significant difference in scoliosis based on defect size, there was a trend toward increased scoliosis with large defects. The failure to demonstrate this difference is likely due to the lack of sensitivity of chart review and clinical examination at detecting mild scoliosis compared to focused radiologic review. We similarly found no difference among chest wall deformities when comparing flap to patch repair, and while an abdominal wall bulge is often present after flap repair it improves over time as the single layer external oblique portion of the abdominal wall reconstruction strengthens.

Large diaphragmatic hernias are technically challenging to repair. In a review of the national CDH database, 88% of large defects are repaired using a synthetic patch [1]. Our preference has been to utilize a split abdominal wall muscle flap when primary repair cannot be achieved. This technique has the advantage of a tension-free repair with vascularized, innervated, autologous tissue that will grow with the patient, and avoid infection. We have seen significantly fewer recurrences using a split abdominal wall muscle flap compared to a synthetic patch repair [9], and the muscle flap repair can be performed safely in the setting of extracorporeal membrane oxygenation [25]. When presenting our cohort of patients, the question arises as to whether this type of repair increases the risk of chest wall deformity. Based on our results, split abdominal wall muscle flap does not increase the risk of musculoskeletal deformity compared to patch repair.

There are several limitations to our study. First and foremost is that a portion of this study was done by retrospective review. We attempted to assess the accuracy of this review with focused prospective clinic visits in a subset of children. All patients with large defects were invited to participate in the prospective follow-up but only 26% participated. We are unable to determine whether this is truly representative or if there was a self-selection bias. Secondly, there has been a trend towards flap repair at our institution, and

because of this the number of patch repairs is limited. It is also not possible to clearly define why each surgeon chose one operation over the other. Additionally, the follow-up of these CDH patients is ongoing. Skeletal deformities, especially pectus deformities, are not typically managed until teenage years and 60% of these patients have not yet reached the age of 13. We assume that most of these children would seek help for skeletal anomalies at our hospital or an affiliated Intermountain Healthcare facility, but it is possible that some patients have been lost to follow-up.

4. Conclusion

We found a high incidence of spinal and chest wall deformities in patients following repair of a large CDH. This does not seem to be dependent on whether a split abdominal wall muscle flap or patch repair was performed. Though these deformities are increased, they very rarely have any therapeutic consequences. Whether chest wall deformities result from the repair or are part of the somatic constellation associated with large CDH defects remains unclear.

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