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Long-term burden of care and radiation exposure in survivors of esophageal atresia

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ABSTRACT

Background: Patients with esophageal atresia with or without tracheoesophageal fistula (EA/TEF) historically have had a high risk of neonatal mortality but the majority of patients are now expected to live into adulthood. However, the long-term burden of care among recent EA/TEF survivors has not been documented.
Methods: A single-institution retrospective review of newborns with EA/TEF treated from 2001-2005 was conducted, including initial and total hospitalization length of stay, and number of clinic visits and procedures requiring general anesthesia in the first three years of life. Exposure to and number of radiological studies involving ionizing radiation (IR) were recorded.
Results: Seventy-one of 78 (91%) patients survived to discharge and 69 were included for analysis. Mean length of initial hospital stay was 51.3 (range 9-390) days. By age 3 years, patients required 4.5 (mean, range 1-23) procedures performed under general anesthesia, attended 13.5 (mean, range 3-40) outpatient visits and were exposed to 17.4 mSv (mean, range 3.0-59.9) of IR from 40 (mean, range 5-165) radiological studies.
Conclusion: Patients with EA/TEF need complex and frequent hospital-based care from infancy to early childhood. Opportunities to critically review clinical services and imaging needs should be explored to improve the experience of patients and their families.

1. Background

Esophageal atresia with or without tracheoesophageal fistula (EA/TEF) is a congenital anomaly of the foregut affecting 1 in 3000 live births [1]. With improved surgical and neonatal care, survival has reached over 90% [2]. However, patients with EA/TEF may experience esophageal and respiratory morbidities throughout their childhood, including recurrent fistulas, esophageal strictures, chronic dysphagia, gastroesophageal reflux, pulmonary infections, asthma, and tracheomalacia [3]. Some may have associated genetic syndromes, such as Trisomy 21 [4]. Furthermore, EA/TEF constitutes part of the spectrum of VACTERL association – vertebral, anorectal, cardiac, tracheoesophageal, renal, and limb anomalies that commonly co-occur, adding to the complexity of their clinical management [5].

Diagnostic imaging studies and interventional radiology procedures are important aspects of EA/TEF primary and follow-up care. These include imaging modalities that utilize ionizing radiation (IR), such as radiographs, fluoroscopy, computed tomography (CT) and nuclear medicine, and those that do not utilize IR, such as ultrasound and magnetic resonance imaging (MRI). Varying speciality outpatient clinics and complex hospitalizations mean that the long term burden of care is considerable. However, studies quantifying both clinical care burden and cumulative radiation exposure for patients with EA/TEF are lacking currently. Our aim was to characterize the burden of care faced by a five year cohort of patients with EA/TEF during primary and follow-up care at our institution.

2. Methods

With institutional ethics review board approval (REB #1000032265), a retrospective review of patients born from 2001-2005 and treated for EA/TEF at our tertiary care pediatric hospital was conducted. Patients who did not survive to discharge and those whose geographical location precluded regular follow-up at our institution were excluded from analysis. The study cohort was analyzed for demographic characteristics, hospital length of stay during their primary repair, total length of hospital stay in the first three years of life, number of emergency department (ED) visits, procedures performed under general anesthesia (GA), number of and interval between scheduled outpatient clinic visits, and radiation exposure during the first three years of life.

For the diagnosis of esophageal atresia, radiography of the chest +/- abdomen was obtained for all newborns, followed by VACTERL workup including radiography of the spine, echocardiography, and kidney

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ultrasound. All other radiological investigations were obtained at the discretion of the treating physicians.

Institutional Picture Archiving and Communication System and interventional radiology databases were used to determine the frequency of diagnostic imaging studies and interventional radiology procedures performed on patients in their first three years of life and to assign appropriate radiation dose estimates. Frequency and radiation dose were estimated for the following imaging modalities which utilize IR: radiography, diagnostic fluoroscopy, interventional radiology, computed tomography, nuclear medicine, and cardiac catheterization. Frequency data were collected for imaging modalities which do not utilize IR: ultrasound and magnetic resonance imaging.

Radiation dose data were calculated as Effective Dose estimates per examination in units of millisieverts (mSv) and then summated per patient as Cumulative Effective Dose (CED). Factors considered in dose assignment included: type of study, patient age, number of views (radiography), number of scanning phases and coverage (CT), fluoroscopy time and number of digital subtraction angiograph frames (interventional radiology), and changes to institutional diagnostic equipment that resulted in radiation dose reductions during the study period (e.g. the introduction of pulsed fluoroscopy capability). For the majority of studies and procedures, dose assignment was based on institutional data applicable to the time period of our study [6–10]. Published values from the pediatric radiology literature [11–19] were used for the small number of examinations for which institutional data were not available. Table 1 summarizes the Effective Dose estimates for common radiological studies and procedures. Diagnostic images supplied by referring hospitals were included in CED calculations and assigned the same dose as studies performed at our institution.

Table 1
Effective dose estimates for common radiological studies and procedures during the study period, 2001–2008.

Type of Examination	Patient Age	
	0–3 months	4 months–2 years 11 months
Plain radiography, mSv per view		
Chest	0.01	0.01
Abdomen	0.017	0.04
Chest + abdomen	0.035	0.035
Pelvis	0.01	0.015
Spine (cervical, thoracic or lumbar)	0.04	0.04
Extremity	0.0001	0.0001
Skull	0.015	0.015
Skeletal survey	0.22	0.23
CT, mSv per study		
Head, 1 phase (with or without contrast)	4.2	3.6
Head, 2 phases (with and without contrast)	8.4	7.2
Petrous bones	4.1	2.6
Chest	2.8	3.4
Abdomen and pelvis	6.5	5.5
Diagnostic fluoroscopy, mSv per study (years 2001–2005)[a]		
Upper gastrointestinal series	3.6	2.7
Esophagram	3.6	2.7
Voiding cystourethrogram	0.36	0.42
Feeding Study	1.05	0.6
NJ tube insertion	3.0	1.8
NG tube insertion	1.5	0.9
Nuclear medicine, mSv per study		
DMSA renal scan	1.9	1.9
Mag 3 renal scan	0.7	0.7
Bone scan	7.0	7.0
Bone mineral densitometry	0.0005	0.0005
HIDA scan	5.2	5.2
Lung perfusion scan	1.1	1.1
Gastric emptying study	2.2	2.2
Cardiac catheterization, mSv per study		
Diagnostic or interventional cardiac Catheterization[b]	6.0	4.0
Interventional radiology, mSv per minute (from May 2001 to the end of study)[c]		Age 0–3 years
PICC line insertion		0.035
CVL insertion		0.035
Esophageal dilatation		0.035
Pleural drainage		0.035
G tube insertion		0.156
G tube maintenance		0.139
GJ tube insertion		0.156
GJ tube maintenance		0.145

[a] 2006–end of study, divide dose by factor of 3 (with the introduction of pulse fluoroscopy).
[b] Dose estimate from literature review.
[c] January–April 2001, multiply dose by factor of 2.

3. Results

There were 78 neonates with EA/TEF treated at our institution from 2001 to 2005 (Table 2). Of these, 71 (91%) survived to discharge. Causes of death included sepsis, liver failure, pulmonary hypoplasia, gastric perforation, air leak with ventilator failure, and extensive small bowel necrosis. Two patients were excluded from analysis due to geographical location precluding regular follow up. Of the 69 patients included for burden of care analysis, 32 (46%) were male, 20 (29%) were born preterm (less than 37 weeks gestation), and 19 (28%) were diagnosed with VACTERL association. The majority of patients (55 patients, 80%) had esophageal atresia with a distal tracheoesophageal fistula (Type C), while only 3 patients (4%) had long-gap esophageal atresia.

Patients had a mean length of stay of 51 (median 30, range 9–390) days during their initial hospitalization, with EA/TEF repair usually performed in the first few days of life. Thirty patients (43%) required tube feeds via gastrostomy tube. By age 3 years, patients required on average 4 (median 3, range 1–23) procedures performed under general anesthesia, including surgical procedures such as fundoplication, interventional radiology procedures such as esophageal dilatation and gastrostomy tube placement, and diagnostic studies such as CT and MRI studies. Patients also had on average 3 (median 1, range 0–19) ED visits. Seventy percent of procedures performed under GA and 48% of ED visits took place in the first year of life.

All patients who survived to discharge were seen as outpatients at the general surgery clinic one month after discharge, followed by semi-annual or annual visits if asymptomatic. If symptomatic, they were referred to the appropriate medical or surgical services. In our study, patients visited the hospital on average 13 (median 11, range 3–40) times for outpatient clinic visits by age 3 years. Furthermore, they attended two outpatient clinics within two weeks of each other on average 2 (median 1, range 0–14) times.

Repeat hospitalizations before the age of 3 years were common. Of the 69 patients in the cohort, 32 (46%) were admitted at least once from the ED, while 28 (41%) patients were admitted to hospital at least once by another medical service. Including initial admission and subsequent readmissions, patients required 3 hospitalizations (mean, median 2, range 1–21) in the first three years of life, resulting in total hospital length of stay of 74 days (mean, median 43, range 9–391) (Table 3).

In the first three years of life, the mean IR exposure for the EA/TEF survivor was 17.4 mSv (median 14.8, range 3.0–59.9 mSv) from an average of 40 (median 24, range 5–165) radiological studies (Table 4). The largest contributor to the overall number of radiological studies involving IR was radiography (i.e. chest and abdominal radiographs), with a mean of 23 radiographs per patient (range 2–129), and 57.5% of the total number of radiological studies. However, radiography only accounted for a mean of 0.6 mSv (range 0.03–3.3), or 3%, of IR exposure per patient. Diagnostic fluoroscopy (i.e. esophagrams) was the modality with the largest effect on IR exposure, contributing to 70% of the total CED with 12.2 mSv (mean, range 2.5–42.3) from 5 (mean, range 1–19) studies per patient (12.5% of studies). At least one of the 5 diagnostic fluoroscopy studies was a routine post-repair esophagram