

Urinary Tract and Other Associated Anomalies in Newborns With Esophageal Atresia

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Esophageal atresia is often associated with other anomalies. Hereditary and environmental factors may influence the incidence of associated anomalies, particularly of the urogenital system. We had 63 neonates with esophageal atresia admitted to 2 centers in Hamadan, Iran, from 2002 to 2008. They were 38 girls (60.3%) and 25 boy (39.7%). Tracheoesophageal fistula was present in 54 neonates (85.7%), and other associated anomalies in 10 (15.9%). Cardiac anomalies were found in 7 neonates; anorectal anomalies, in 4; urinary tract anomalies, in 2; and limb anomaly, in 1. Urinary tract anomalies (3.2%) were bilateral polycystic kidney in 1 neonate and unilateral hydronephrosis due to ureteropelvic junction obstruction in another. Both neonates with urinary tract anomalies were female and both had tracheoesophageal fistula, as well. Many of the associated congenital abnormalities influence the management protocol of esophageal atresia, and therefore, should be detected as soon as possible after birth.

Keywords: esophageal atresia, newborn infant, urinary tract, congenital abnormalities

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Esophageal atresia can occur as an isolated entity or in association with one or more fistulas communicating between the abnormal esophagus and the trachea. The embryologic missteps leading to esophageal atresia are associated with varying components of the malformation constellation termed VACTERL, which refers to anomalies of the vertebrae (V), atresias in the gastrointestinal tract (A), congenital heart lesions (C), tracheoesophageal defects (TE), renal and distal urinary tract anomalies (R), and limb lesions (L).⁽¹⁾ If any of these anomalies are present, the presence of the others must be assessed.⁽²⁾ Thus, urinary tract abnormalities must be considered in all of neonates with esophageal atresia with or without tracheoesophageal

fistula.^(1,3,4) Early ultrasonographic assessment is mandatory and useful to evaluate associated kidney or ureteral anomalies, or both.^(1,3-5) In addition, the kidneys may be palpable on physical examination.^(3,5) Early recognition of newborns with no prospect of long-term survival (bilateral renal agenesis) will avoid unnecessary surgery on the esophagus.^(1,3-5)

The reported incidence of urinary tract anomalies associated with esophageal atresia has been varying between 4% and 64% in different studies.^(3,6,7) Hereditary and environmental factors may influence the incidence of associated anomalies, particularly of the urogenital system.⁽⁵⁻⁸⁾ The initial reports documenting this spectrum of associated defects appeared over

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30 years earlier, and the incidence of urinary tract and other associated anomalies has not been precisely quantified in recently.^(5,9) Multiple studies have shown that knowledge on the diagnosis of concomitant anomalies with esophageal atresia, especially urinary tract abnormalities, can improve prognosis of the patients.⁽⁵⁻¹⁰⁾ We reviewed our hospital records for evaluation of the prevalence of urinary tract and other associated anomalies with esophageal atresia in neonates.

This study was a retrospective investigation on hospitalized neonates at Ekbatan and Besat hospitals of Hamadan University of Medical Sciences. Neonates with esophageal atresia who had been admitted between 2002 and 2008 were identified. Their documents were evaluated and data including age, sex, birth weight, diagnosed urinary tract anomalies, and other associated anomalies were collected. All of these neonates had drooling or excessive salivation and respiratory difficulties in the first few days of life. Passing of a nasogastric tube had not been possible. In attempts to pass a nasogastric tube, chest radiography had shown the tube curling up in the upper esophageal pouch. The diagnosis of isolated esophageal atresia with no tracheoesophageal fistula had been made in case of no air in the gastrointestinal tract on chest radiography.

In our two centers, the diagnostic management plan for evaluation of associated congenital anomalies had included the following: lateral lumbar radiography, for vertebral anomalies (multiple or single hemivertebrae, scoliosis, and rib deformities); rectal examination, for imperforate anus and anorectal anomalies; echocardiography, for congenital heart lesions (ventricular septal defects, atrial septal defects, and tetralogy of Fallot); posteroanterior and lateral plain chest radiographies with nasogastric tube opaque line, for tracheoesophageal fistula and esophageal atresia; urinary tract ultrasonography, for urinary tract anomalies (renal agenesis, Potter syndrome, bilateral renal agenesis or dysplasia, horseshoe kidney, polycystic kidneys, urethral atresia, and ureteral malformations); and limb radiography for limb anomalies (radial dysplasia,

absent radius, syndactyly, polydactyly, and tibial deformities). Also we had performed voiding cystourethrography and technetium Tc 99m diethylenetriamine pentaacetic acid renal scintigraphy for the patients suspected to have urinary tract anomalies.

We found that during the 6-year studied period, 63 neonates had been diagnosed with esophageal atresia. A female predominance was seen in these neonates; they were 38 girls (60.3%) and 25 boys (39.7%). The mean age of the neonates was 2.33 ± 1.90 days (range, 1 to 11 days) at diagnosis, and their mean birth weight was 2678.6 ± 511.3 g (range, 1350 g to 3600 g). Tracheoesophageal fistula was present in 54 neonates (85.7%).

Ten neonates (15.9%) had other anomalies (regardless of having tracheoesophageal fistula or not), including cardiac anomalies in 7 neonates (ventricular septal defects and atrial septal defects), anorectal anomalies in 4 (imperforated anus), urinary tract anomalies in 2, and limb anomaly in 1 (absent radius). Urinary tract anomalies were bilateral polycystic kidney in 1 neonate and unilateral hydronephrosis due to ureteropelvic junction obstruction in another. Both neonates with urinary tract anomalies were female and both had tracheoesophageal fistula. In one of these patients, voiding cystourethrography revealed vesicoureteral reflux with unilateral hydronephrosis and technetium Tc 99m diethylenetriamine pentaacetic acid renal scan showed ureteropelvic junction obstruction. Both of the neonates had been referred to our hospital within the first 24 hours after birth. Their weights were 3300 g and 2850 g. Their laboratory tests and vital signs were normal at admission. Esophagoplasty and fistulography had been performed for both of the newborns. Five days after the operation, esophagography revealed no leakage or abnormality. After discharge, they were referred to a pediatric nephrologist for follow-up.

Esophageal atresia is a life-threatening malformation with unknown pathogenesis. Management of acute and chronic problems related to the associated anomalies, especially those of the urinary tract, are crucial, because they are the main causes of death in newborns

with esophageal atresia.^(1-3,11) On the other hand, early recognition of urinary tract anomalies with no prospect of long-term survival (eg, bilateral renal agenesis) will avoid unnecessary surgeries.^(9,10,12) In our study, 63 neonates with esophageal atresia referred to our centers during a 6-year period were evaluated for urinary tract and other associated anomalies. Urinary tract anomalies were found in 3.2% of the patients who had bilateral polycystic kidney and unilateral hydronephrosis due to ureteropelvic junction obstruction. Similar anomalies have been described in other studies on esophageal atresia.⁽³⁻⁷⁾

The frequency of tracheoesophageal fistula in our study was 85.7%, which was consistent with the reported 80% to 90% incidence rates of tracheoesophageal fistula in patients with esophageal atresia.⁽¹⁻¹¹⁾ In our study, 15.9% of the infants had been affected by other anomalies (with or without tracheoesophageal fistula). Previous studies have shown a 40% to 57% incidence of associated anomalies.^(2-7,9,11) Chittmitrapap and colleagues reviewed their 8-year data and found that 48% of their patients with esophageal atresia had associated anomalies, as well.⁽⁶⁾ Van Heurn and colleagues' study compared the associated anomalies in neonates with esophageal atresia was between the Asian and European neonates. There were no significant differences in the incidence of cardiovascular, anorectal, and musculoskeletal anomalies between the two populations. However, the European patients had a significantly higher incidence of urinary tract anomalies (26% versus 4%, $P = .006$).⁽⁷⁾

The frequency of urinary tract anomalies has been varying in different reports. Brown and colleagues documented a renal anomaly incidence of 16% in infants with esophageal atresia and tracheoesophageal fistula,⁽⁹⁾ while Muraji and Mahour reported that 9 of 14 infants with esophageal atresia with or without tracheoesophageal fistula had urinary tract anomalies.⁽¹⁰⁾ Gunn and coworkers found urinary tract abnormalities is 14.3 per 1000 births with esophageal atresia.⁽¹³⁾ Overall, it has been shown that urinary tract abnormalities are the second most common associated defects with esophageal atresia after cardiac abnormalities.⁽¹⁻⁵⁾

Hereditary factors may influence the incidence of associated anomalies in neonates with esophageal atresia, particularly of the urinary tract anomalies. However, environmental factors cannot be excluded.^(6,7,10) Significant discrepancies in reported incidence of urinary tract anomalies in previous studies warrants studies on larger samples with special attention to the genetic and environmental causal factors. Many of the associated congenital abnormalities influence the way in which the esophageal atresia is managed, and therefore, should be detected as soon as possible after birth. Thus, knowledge on the local epidemiology and etiologies is crucial.

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CONFLICT OF INTEREST

None declared.

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