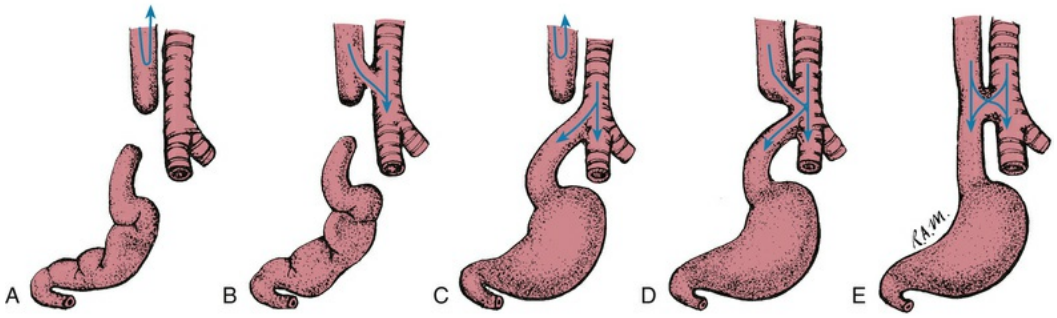

FIG 22-5 **A** to **E,** Five most common types of esophageal atresia (EA) and tracheoesophageal fistula (TEF).

The incidence of EA is estimated to be approximately 1 in 4000 live births (Kunisaki and Foker, 2012). There appears to be a slightly higher incidence in males, and the birth weight of most affected infants is significantly lower than average, with an unusually high incidence of preterm birth with EA and a subsequent increase in mortality. A history of maternal polyhydramnios is common.

Approximately 50% of the cases of EA/TEF are a component of VATER or VACTERL association, which are acronyms used to describe associated anomalies (VATER for *v*ertebral defects, imperforate *a*nus, *t*racheo*e*sophageal fistula, and *r*adial and *r*enal dysplasia; and VACTERL for *v*ertebral, *a*nal, *c*ardiac, *t*racheal, *e*sophageal, *r*enal, and *l*imb) (Khan and Orenstein, 2016b). Cardiac anomalies may also occur with EA/TEF; therefore, all patients should undergo a workup for associated anomalies.

Pathophysiology

Anomalies involving the trachea and esophagus are caused by defective separation, incomplete fusion of the tracheal folds after this separation, or altered cellular growth during embryonic development. In the most frequently encountered form of EA and TEF (80% to 90% of cases), the proximal esophageal segment terminates in a blind pouch, and the distal segment is connected to the trachea or primary bronchus by a short fistula at or near the bifurcation (see Fig. 22-5, *C*). The second most common variety (7% to 8%) consists of a blind pouch at each end, widely separated and with no communication to the trachea (see Fig. 22-5, *A*). An H-type EA refers to an otherwise normal trachea and esophagus connected by a fistula (4% to 5%) (see Fig. 22-5, *E*). Extremely rare anomalies