Table 10-1 Cause of Death Related with Age

Causes*	Rate†
Younger than 1 year	**660.6**
Congenital malformations, deformations, and chromosomal anomalies	
Disorders related to short gestation and low birth weight	
Sudden infant death syndrome (SIDS)	
Newborn affected by maternal complications of pregnancy	
Accidents (unintentional injuries)	
Newborn affected by complications of placenta, cord, and membranes	
Bacterial sepsis of newborn	
Respiratory distress of newborn	
Diseases of the circulatory system	
Neonatal hemorrhage	
1-4 Years	**28.3**
Accidents (unintentional injuries)	
Congenital malformations, deformations, and chromosomal abnormalities	
Assault (homicide)	
Malignant neoplasms	
Diseases of the heart‡	
5-9 Years	**12.5**
Accidents (unintentional injuries)	
Malignant neoplasms	
Congenital malformations, deformations, and chromosomal abnormalities	
Assault (homicide)	
Influenza and pneumonia	
10-14 Years	**15.7**
Accidents (unintentional injuries)	
Malignant neoplasms	
Intentional self-harm (suicide)	
Assault (homicide)	
Congenital malformations, deformations, and chromosomal anomalies	

*Causes are listed in decreasing order of frequency. All causes and rates are based on 2008 (final) and 2009 (preliminary) data.
†Rates are expressed per 100,000 population from all causes within each age group.
‡Excludes congenital heart disease.
Data source: Centers for Disease Control and Prevention/NCHS, National Vital Statistics System: mortality, 2009 and 2008 (www.cdc.gov/nchs/nvss/mortality_tables.htm).

Congenital Anomalies

Congenital anomalies are anatomic defects that are present at birth, but some, such as cardiac defects and renal anomalies, may not become clinically apparent until years later. The term *congenital* means "born with," but it does not imply or exclude a genetic basis for the birth defect. It is estimated that about 120,000 (1 in 33) babies are born with a birth defect each year in the United States. They are the most common cause of mortality in the first year and contribute significantly to morbidity and mortality throughout the early years of life. In a sense, anomalies found in live-born infants represent the less serious developmental failures in embryogenesis that are compatible with live birth. Perhaps 20% of fertilized ova are so anomalous that they are blighted from the outset. Others may be compatible with early fetal development, only to lead to spontaneous abortion. Less severe anomalies allow more prolonged intrauterine survival, with some disorders terminating in stillbirth and those still less significant permitting live birth despite the handicaps imposed.

Definitions

The process of morphogenesis (organ and tissue development) can be impaired by a variety of different errors.

- *Malformations* represent primary errors of morphogenesis, in which there is an *intrinsically abnormal developmental process* (Fig. 10-1). Malformations can be the result of a single gene or chromosomal defect, but are more commonly multifactorial in origin. Malformations may present in several patterns. Some, such as congenital heart defects and anencephaly (absence of the brain), involve single body systems, whereas in other cases multiple malformations involving many organs may coexist.
- *Disruptions* result from secondary destruction of an organ or body region that was previously normal in development; thus, in contrast to malformations, disruptions arise from an *extrinsic disturbance in morphogenesis*. *Amniotic bands*, denoting rupture of amnion with resultant formation of "bands" that encircle, compress, or attach to parts of the developing fetus, are the classic example of a disruption (Fig. 10-2). A variety of environmental agents may cause disruptions (see later). Understandably, disruptions are not heritable and hence are not associated with risk of recurrence in subsequent pregnancies.
- *Deformations*, like disruptions, also represent an *extrinsic disturbance of development* rather than an intrinsic error of morphogenesis. Deformations are common problems, affecting approximately 2% of newborn infants to varying degrees. Fundamental to the pathogenesis of deformations is localized or generalized compression of the growing fetus by *abnormal biomechanical forces*, leading eventually to a variety of structural abnormalities. The most common underlying factor responsible for deformations is *uterine constraint*. Between the thirty-fifth and thirty-eighth weeks of gestation, rapid increase in the size of the fetus outpaces the growth of the uterus, and the relative amount of amniotic fluid (which normally acts as a cushion) also decreases. Thus, even the normal fetus is subjected to some form of uterine constraint. Several factors increase the likelihood of excessive compression of the fetus resulting in deformations. *Maternal factors* include first pregnancy, small uterus, malformed (bicornuate) uterus, and leiomyomas. *Fetal or placental factors* include oligohydramnios, multiple fetuses, and abnormal fetal presentation. An example of a deformation is clubfeet, often a component of Potter sequence, described later.
- A *sequence* is a cascade of anomalies triggered by one initiating aberration. Approximately half the time, congenital anomalies occur singly; in the remaining cases, multiple congenital anomalies are recognized. In some instances the constellation of anomalies may be explained by a single, localized aberration in organogenesis (malformation, disruption, or deformation) that

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