for the standardization of respiratory care for patients with SMA have been published elsewhere (Schroth, 2009).

Prognosis

Prognosis varies according to the age of onset or group as described in Box 30-7. Individuals with SMA type 1 may succumb to respiratory infections or failure between 1 and 24 months of age (Iannaccone and Burghes, 2002; Sarnat, 2016a); however, some may live into their third or fourth decade of life. A significant number of infants with SMA require a tracheotomy, and associated medical conditions in survivors include gastroesophageal reflux, scoliosis, early onset puberty, hip dysplasia, and recurrent oral candidiasis (Bach, 2007). Drug therapy with riluzole, valproic acid, gabapentin, and oral phenylbutyrate has been shown to slow the progression of the condition, but none has demonstrated significant overall benefits (Wadman, Bosboom, van der Pol, et al, 2012; Sarnat, 2016a).

Nursing Care Management

An infant or small child with progressive muscle weakness requires nursing care similar to that of an immobilized patient (see Chapter 29). However, the underlying goal of treatment should be to assist the child and family in dealing with the illness while progressing toward a life of normalization within the child's capabilities. Special attention should be directed to preventing muscle and joint contractures, promoting independence in performance of ADLs, and becoming incorporated into the mainstream of school when possible. In addition, parents need support and resources to be able to provide for the child and remain an intact family. Because children with neuromuscular disease have abnormal breathing patterns that often contribute to early death, it is important to assess adequate oxygenation, especially during the sleep phase when shallow breathing occurs and hypoxemia may develop. Home pulse oximetry may be used to assess the child during sleep and provide noninvasive mechanical ventilation as necessary (Bush, Fraser, Jardine, et al, 2005; Young, Lowe, Fitzgerald, et al, 2007) (see Duchenne [Pseudohypertrophic] Muscular Dystrophy later in this chapter for respiratory management). Supportive care also includes management of orthoses and other orthopedic equipment as required. Because children with SMA are intellectually normal,