The signs of bilirubin encephalopathy are those of CNS depression or excitation. Prodromal symptoms consist of decreased activity, lethargy, irritability, hypotonia, and seizures. Later these subtle findings are followed by development of athetoid cerebral palsy, gaze palsies, and deafness (Watson, 2009). Motor skills are delayed, and dental enamel hypoplasia may also occur. Those who survive may eventually show evidence of neurologic damage, such as cognitive delay, ADHD, delayed or abnormal motor movement (especially ataxia or athetosis), behavior disorders, perceptual problems, or sensorineural hearing loss.

Therapeutic Management

The primary goals in the treatment of hyperbilirubinemia are to identify infants at high risk for hyperbilirubinemia; monitor serum bilirubin levels; prevent bilirubin encephalopathy; and, as in any blood group incompatibility, to reverse the hemolytic process. The main form of treatment involves the use of phototherapy. Exchange transfusion is generally used for reducing dangerously high bilirubin levels that may occur with hemolytic disease.

Intravenous immunoglobulin (IVIG) is effective in reducing bilirubin levels in infants with Rh isoimmunization and ABO incompatibility (Watson, 2009) and is recommended by the American Academy of Pediatrics (American Academy of Pediatrics, Subcommittee on Hyperbilirubinemia, 2004). The evidence supporting the use of IVIG is limited and further research is recommended (Keir, Dunn, and Callum, 2013).

Healthy near-term and full-term infants with jaundice may also benefit from early initiation of feedings and frequent breastfeeding. These preventive measures are aimed at promoting increased intestinal motility, decreasing enterohepatic shunting, and establishing normal bacterial flora in the bowel to effectively enhance the excretion of unconjugated bilirubin.

Phototherapy consists of the application of a special source of light (irradiance) to the infant's exposed skin (Fig. 8-17). Light promotes bilirubin excretion by **photoisomerization**, which alters the structure of bilirubin to a soluble form (**lumirubin**) for easier excretion.