Chapter 35

The Special Care Nursery

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This chapter describes the history and organization of the special care nursery and discusses which neonates and infants are at risk for central nervous system (CNS) dysfunction or developmental delay. A theoretic basis for physical therapy examination, evaluation, prognosis, and interventions for infants in the special care nursery is presented and recommendations are provided for clinical practice. The gestational and pathophysiologic conditions considered in this chapter are prematurity, hypoxic-

ischemic encephalopathy, fetal alcohol syndrome, fetal abstinence syndrome, exposure to human immunodeficiency virus (HIV) infection, neonatal seizures, birth injuries related to the CNS, and spina bifida. The follow-up of infants after discharge from the special care nursery is addressed, and two case histories illustrate and integrate the material presented in this chapter.

HISTORY OF THE SPECIAL CARE NURSERY

Modern neonatal care was born with the development of the first incubator by Couveuse in France in 1880 (Hodgman, 1985). The first text on the premature infant, *The Nursling*, authored by Budin, a student of Couveuse, was published in 1900. Dr. Martin Couney, who was one of Budin's students, used these principles of treatment for the premature infant, and in a bizarre entrepreneurial twist, exhibited them to the public for a fee (Silverman, 1979). The main principles of neonatal care were support of body temperature, control of nosocomial infection, minimal handling, and provision of special nursing care. Interestingly, nurseries were quiet, and lights were dimmed at night. Dr. Julius Hess attended this exhibition in Chicago and applied these principles in the late 1940s. Dr. Hess achieved a neonatal mortality rate for preterm infants of 20%, which was respectable for the time. In response to the increased survival rate of premature infants reported by Hess, these principles of care spread across the United States.

During the 1950s, a number of cities developed centers for the care of premature infants and a number of states developed maternal mortality committees that gathered data to be used as a basis for planning activities directed at preventing maternal death. During the 1960s, Arizona, Massachusetts, and Wisconsin promulgated standards for maternity units and developed regional perinatal care centers. Reports from these three states and several professional organizations, including the American Medical Association, the American College of Obstetricians and Gynecologists, the American Academy of Pediatrics, and the Academy of Family Physicians, stimulated the development of the regional organization of perinatal services (Fanaroff & Graven, 1992).

By the late 1960s, full-term infants with health complications were also being treated in the neonatal nursery. Advances in microlaboratory techniques for biochemical determinations from minute quantities of blood and the development of miniaturized monitoring equipment, ventilatory support systems, and means to conserve body heat improved the care of the neonate with serious illness (Fanaroff & Graven, 1992). Expansion of neonatal pharmacology, widespread use of phototherapy for management of hyperbilirubinemia, and methods of delivery of high-caloric solutions parenterally when oral feeding was not possible also improved the chances for survival of the very sick neonate. In 1975, the emergence of the new subspecialties of neonatology and perinatology provided specialists in the field of caring for infants in the high-technology nursery.

During the past two decades there has been considerable progress in the treatment of neonates and children with critical illnesses. For example, there has been an increase in the survival rate of infants born at 23 to 25 weeks of gestational age from 27% in 1984 to 1989 to 42% in 1990 to 1995, with most of the increase in disability being mild (Emsley et al., 1998). The improvement in survival and quality of life for these patients resulted from nationwide development of regional neonatal intensive care units (NICUs) in which an organized, highly specialized, multidisciplinary approach became the standard of care. The number of neonates needing close supervision and expert cardiorespiratory and metabolic support is large enough to make such units an essential component of a perinatal health care delivery system (Sarnaik & Preston, 1985). The increase in the number of NICUs resulted in a significant drop in neonatal and perinatal mortality rates of low birth weight (LBW) infants (Lubchenco et al., 1974; McDonald, 1981; Teberg et al., 1977). By 1996, the survival rate of extremely low birth weight (ELBW) infants (750–1000 g) had increased to 86% and the survival rate of infants who weighed 500 to 750 g at birth was 54%, which are in stark contrast to the 10% survival rate in 1960 for infants with birth weights of 750 to 1000 g (Lemons and Stevenson and Wright, 1998). Currently, there are 700 NICUs in the United States, which is a 30-fold to 40-fold increase since the 1960s. Long-term outcome of the very low birth weight (VLBW) infant, however, came under scrutiny with the suspicion that the typical nursery stay of several weeks may have a significantly detrimental effect on later behavioral performance (Hodgman, 1985). Research ensued on the effects of different sensory input during the NICU stay and the concept of neonatal care facilitating optimal development was born (Als, 1986; Als et al., 1986, 1994; Glass et al., 1985; Hilton, 1987).

ORGANIZATION OF PERINATAL SERVICES

Special care units are designed to meet a wide range of special needs, from the monitoring of apparently well infants at risk of serious illness to the intensive treatment of infants with acute illness. This range of services

Box 35-1 In-Hospital Perinatal Services

Basic Care: Surveillance and care of all patients admitted to obstetric service with triage to identify high-risk patients.

Specialty Care: Care of high-risk mothers and fetuses. Stabilization of ill newborns prior to transfer. Care of preterm infants with birth weight of 1500 g or more.

Subspecialty Care: Provision of comprehensive perinatal services for mothers and neonates of all risk categories. Research and educational support. Analysis and evaluation of regional data. Initial evaluation of new high-risk technologies.

Adapted from American Academy of Pediatrics, American College of Obstetricians and Gynecologists. Guidelines for Perinatal Care. 4th ed. Elk Grove Village, IL, Washington, DC: American Academy of Pediatrics, American College of Obstetricians and Gynecologists; 1997.

requires that the special care nursery be arranged for graduated care to meet the diverse and changing needs of infants (Whaley & Wong, 1991). Because it was not economically feasible for every hospital to have the personnel and technology to care for neonates with complex needs, these services were originally organized on a regional basis. Recent efforts to contain the cost of health care has led to deregionalization in many areas, and blurring of differences in levels of care. Levels of care are currently described by the American Academy of Pediatrics (1997) as Level I Basic Care, Level II Specialty Care, and Level III Subspecialty Care (Box 35-1). A broad-based Committee on Perinatal Health recently concluded that a strong focus of perinatal care is preventive health care, education, and counseling (Bagwell & Armstrong, 1998).

THE NEONATAL INTENSIVE CARE UNIT ENVIRONMENT

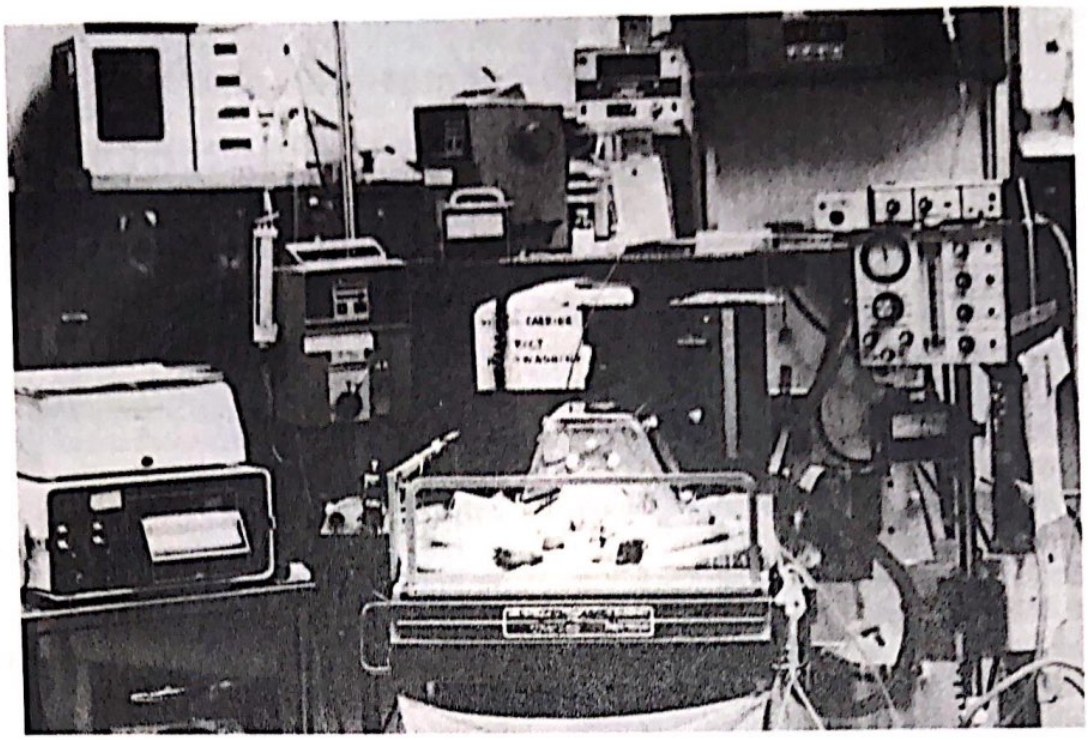
NICUs were designed to decrease neonatal morbidity and mortality rates. In an attempt to meet this goal, the NICU provides the neonate with a habitat for growth starkly different from the intrauterine environment. The NICU is a busy, often crowded place, where the atmosphere is frequently high pressure (Pelletier & Palmeri, 1985). The intrauterine environment is replaced by bright lights, high noise levels, and the intrusive medical procedures characteristic of high-technology treatment (Campbell, 1986; Gottfried, 1985). Each tiny baby lies in an incubator, an open warmer, or a crib, surrounded by and connected to ventilators and monitors, including heart, apnea, and oxygen monitors and infusion pumps (Fig. 35-1). The incubator is often surrounded by equipment including phototherapy lights; diagnostic transilluminators; and portable radiographic, electroencephalographic, and ultrasonographic units. The amount and complexity of the equipment can be overwhelming to parents and families.

Lighting

Typically, the ambient illumination within the NICU consists of daylight and artificial fluorescent lighting (Moseley et al., 1988). The American Academy of Pediatrics recommends a minimum light intensity of 100 foot-candles at the infant's level for adequate visualization by staff (Weibley, 1989). This same level of illumination may contribute to retinopathy of prematurity (ROP) (Glass et al., 1985; Kretzer & Hittner, 1986). No diurnal rhythmicity of light exists in the NICU, which some investigators believe may interfere with the infant's development of normal biologic rhythms. Glass and colleagues (1985) studied the effects of draping sunglass-filtering material over the incubators. They found that there was a significant increase in ROP in the infants in incubators without the filtering material, especially in the infants who weighed less than 1000 g. As a result of this study, an editorial in the journal that published the study recommended immediate diurnal cycling and dimming of lights (Weibley, 1989). Subsequent studies have not replicated these findings (Phelps & Watts, 2001).

Sound

Sound levels within the incubator have been found to be significantly higher than those measured outside (Kent et al., 2002). In the intrauterine environment, auditory stimuli include sound levels at about 85 dB consisting of rhythmic swooshing and bubbling sounds punctuated by the steady pulse of the maternal heartbeat (Gottfried, 1985; Gottfried & Hodgman, 1984). In the NICU the infant is surrounded by noise on a level comparable with that of auto traffic and, at times, heavy machinery. High noise levels are present from trash receptacles, addressograph machines, centrifuges, telephones, and monitor alarms (Hilton, 1987). About 80% of these peak sounds are human related including opening and closing doors, banging the incubator hood, and tearing and opening bags (Chen & Chang, 2001). These harsh sounds can cause some infants to become hypoxic as part of a startle



• **Figure 35-1** Infant surrounded by typical NICU equipment. *(From Crane, L. Physical therapy for the neonate with respiratory disease. In Irwin, S, & Tecklin, JS [Eds.]. Cardiopulmonary Physical Therapy, 2nd ed. St. Louis: Mosby, 1990, p. 400.)*

response (Thomas, 1989). The environment inside the incubator is characterized by continuous white noise. Harsh mechanical noises penetrate clearly, but speech sounds are indistinct and deflected (Newman, 1981). This lack of distinctness and the deflection of speech sounds may have negative effects on later interactive behavior if the infant learns to look away from the speaker to locate him or her. Drugs commonly used in the nursery are known from animal studies to potentiate noise-induced hearing loss (American Academy of Pediatrics, 1997; Perlstein, 1992). The American Academy of Pediatrics has recommended that noise levels be reduced to less than 70 dB, manufacturers of incubators reduce the noise levels of motors below 58 dB, and physicians limit the use of ototoxic drugs in neonates (Peabody & Lewis, 1985; Perlstein, 1992). Auditory evoked potential tests are typically done in every nursery before a neonate is discharged to establish risk for hearing loss. The American Academy of Pediatrics Committee on Environmental Health has issued recommendations for noise in the environment and its effects on the fetus and newborn, including the infant in the special care nursery. Acoustic foam placed in the corners of the incubator and change in staff behavior decreased noise level (Johnson, 2001; Philbin & Gray, 2002).

Medical Procedures

In the NICU, the infant is placed on a flat mattress and is exposed to a dry, cool, air-filled environment. In the uterine environment the 28- to 32-week fetus sleeps 80% of the time. In contrast, premature infants were found to be disturbed an average of 23 times in 24 hours (Altmier et al., 1999). Tactile input often heralds medical or technical events and causes sustained arousal, which exacts a physiologic toll on the infant. Unable to make sense of life-sustaining efforts, the infant begins to respond negatively to touch (Gottfried & Hodgman, 1984).

DEVELOPMENTAL CARE AND THE SPECIAL CARE NURSERY

In view of the research cited in the preceding sections about the iatrogenic effects of neonatal intensive care, and the shifting of emphasis from survival to the prevention and amelioration of the complications of prematurity, Als and co-workers (1986) have developed the concept of individualized, comprehensive, family-focused, developmentally supportive care for infants in the special care nursery. A growing body of literature supports this type of care (Als, 1986; Als et al., 1986, 1994; Buehler et al., 1995; Byers, 2003; Fleisher et al., 1995; Mouradian & Als, 1994; Parker et al., 1992). Whether or not this specific approach is used, more special care nursery staff members are incorporating a developmentally supportive environment and interventions such as diurnal light cycles; clustering care; specific rest time;

interventions, including sponge baths, as needed rather than on a schedule; skin-to-skin contact, including "kangaroo" care; presentation of organizing environmental input such as music; the scent of the mother on clothing; odor of milk on the pacifier; single room care; and co-bedding of multiple-birth neonates (Bosque et al., 1995; delEstard & Lennox, 1995; Standley & Moore, 1995; Tessier et al., 1998). Research on these methods has shown improvement in such dependent variables as weight gain, days on ventilator, oxygen saturation, number of days in the hospital, infant state, and neuromotor behavior, nonnutritive sucking during gavage feeding, perceptual and cognitive abilities, and parenting process (Brown & Taquino, 2001; Bingham et al., 2003, delEstard & Lennox, 1995; Feldman et al., 2002; Mouradian & Als, 1994; Mueller, 1996).

DEFINING THE AT-RISK INFANT

The meaning of the designation "high-risk infant" differs according to the area of expertise of the professional using the term. The neonatologist defines *risk* as related to morbidity or mortality, whereas psychologists, physical therapists, occupational therapists, and speech therapists may define the *at-risk infant* as one who has a high probability of manifesting developmental delay as a result of exposure to any one of a number of medical factors (Rossetti, 1986; Wilhelm, 1991). High-risk infants are classified according to birth weight, gestational age, and pathophysiologic problems (Whaley & Wong, 1991). Problems related to physiologic status are closely associated with the maturity of the infant and hypoxic episodes during the perinatal period, as well as with fetal exposure to alcohol and drugs and with HIV infection and environmental factors.

PREMATURITY AND LOW BIRTH WEIGHT

Infants born prematurely or who are small for gestational age (SGA) are divided into three major categories: LBW, from 1501 to 2500 g; VLBW, below 1501 g; and ELBW, below 1000 g. Preterm delivery occurs in 8% to 10% of all live births in the United States in spite of current therapies to halt and prevent such deliveries (Harmon & Kenner, 1998). Approximately 75% to 80% of all neonatal morbidity and deaths is due to premature birth. More than 40,000 VLBW infants are born each year, and half of neonatal deaths occur in VLBW infants (Semmler, 1989). These high-risk infants are a heterogeneous group, including infants born preterm (less than 37 weeks of gestation) and those born at term but of reduced weight (Kleigman, 1992).

Black mothers are two to three times more likely than white mothers to deliver VLBW infants (Iyasu & Tomashek, 2002). Causes for LBW and preterm delivery for blacks include racial differences in maternal medical care, stress, and lack of social support. VLBW infants have an increased incidence of neurologic sequelae, delayed development, and lower intellectual and language abilities (Teberg, 1977; Volpe, 1995).

There have been several optimistic reports of prematurity prevention programs that include assessment of prenatal risk, weekly educational interventions, enhanced nutritional support, referral to a perinatologist when necessary, and pH self-measurements. These programs have shown a reduction in NICU admissions and preterm deliveries (Fangman et al., 1994; Joffe et al., 1995; Novy et al., 1995; Saling, 1997).

SGA infants are those whose birth weights are below the 10th percentile of published norms or whose ponderal index (ratio of birth weight to the cube root of the infant's length at birth) is low (Als et al., 1976). Term SGA infants demonstrate developmental problems such as behavioral and learning disorders, and preterm SGA infants may have an even greater prevalence of abnormal developmental problems than term SGA neonates (Kahn-D'Angelo, 1987).

The Clinical Assessment of Gestational Age in the Newborn Infant, developed by Dubowitz and colleagues (1970), is the test most often used by physicians and nurses to assess gestational age. The determination of gestational age is crucial for infants in the special care nursery to interpret neurologic and behavioral findings relative to the correct gestational age. This test includes 10 neurologic and reflex items and 11 external or superficial criteria that are scored on a four-point scale. The accuracy of gestational age is determined within 2 weeks on each assessment with a 95% confidence level. This test was standardized on 167 infants whose mothers were sure of the date of their last menstrual period. It has been used in the assessment of growth in early infancy and, in conjunction with the neurologic examination, to assess differences in development between twins, preterm infants with intraventricular hemorrhage (IVH) and the effects of eclampsia, placental abruption, and intrauterine growth retardation (Francis et al., 1987). Ballard and associates (1991) developed a simplified Dubowitz Newborn Maturity Rating to assess neonates from 20 to 44 weeks of age. Gestational age is also determined by ultrasound, measurements, and amniotic fluid analysis (Kenner &

Lott, 2003). Although gestational age is usually determined by physicians or nurses, the physical therapist should be familiar with the Clinical Assessment of Gestational Age in the Newborn Infant and how gestational age is determined.

MEDICAL COMPLICATIONS AND TREATMENT IN PREMATURITY

PULMONARY COMPLICATIONS

Respiratory Distress Syndrome

Respiratory distress syndrome (RDS), or hyaline membrane disease, is the most common single cause of respiratory distress in neonates. The principal factors in the pathophysiology of RDS are pulmonary immaturity and deficiency of surfactant. Prematurity, low birth weight, low Apgar score at 1 and 5 minutes, maternal age over 34 years, and neonatal transport have been reported as risk factors for RDS (Rubaltelli et al., 1998; Shlossman et al., 1997). Low surfactant production results in increased surface tension, alveolar collapse, diffuse atelectasis, and decreased lung compliance. These factors cause an increase in pulmonary artery pressure that leads to extrapulmonary right-to-left shunting of blood and ventilation-perfusion mismatching (Carlo & Chatburn, 1988; Sweeney & Swanson, 2001; Walsh et al., 1988). Infants with RDS also demonstrate higher heart rates and reduced heart rate variability compared with full-term neonates, indicating that premature birth has an influence on cardiac function for up to 6 months after birth (Henslee et al., 1997).

Prophylactic use of corticosteroids such as dexamethasone and hydrocortisone to accelerate lung development prior to preterm birth are effective in preventing RDS and neonatal death (Crowley, 2002). Intervention for infants with RDS depends on the severity of the disorder and includes oxygen supplementation, assisted ventilation, surfactant administration, and extracorporeal membrane oxygenation (ECMO). Prophylactic surfactant administration of intubated infants younger than 30 weeks of gestational age correlated with an initial improvement in respiratory status and a decrease in the incidence of RDS, pneumothorax, bronchopulmonary dysplasia (BPD), and death (Soll, 2002). Early administration of multiple doses of natural or synthetic surfactant extract results in improved clinical outcome and appears to be the most effective method of administration. When a choice of natural or synthetic surfactant is available, natural surfactant shows greater early improvement in requirement for ventilatory support (Soll, 2002; Yost & Soll, 2002). Continuous distending-pressure ventilators, either positive or negative pressure, are used in the management of RDS. The positive-pressure ventilator is used more often; the negative-pressure ventilator assists ventilation by creating a negative pressure around the thorax and abdomen. Nasal and nasopharyngeal prongs are used for these ventilators (Crane, 1990). (See Chapter 26 for a more detailed description of ventilators.)

Permissive hypercapnia is an experimental ventilatory strategy for acute respiratory failure in which lungs are ventilated with a low inspiratory volume and pressure with the goal being minimization of lung damage during mechanical ventilation (Bigatello et al, 2001). Partial liquid ventilation or perfluorocarbon-associated gas exchange has been used experimentally in conjunction with surfactant administration and has resulted in improved clinical course in a small sample of infants with severe respiratory distress syndrome (Leach et al., 1996). It has been suggested that this technology will provide a strong addition to available treatments for preterm infants and that there will be a resultant decrease in barotrauma and in BPD (Donovan et al., 1998; Vals-I-Soler et al., 2001).

The prognosis of infants with RDS varies with the severity of the original disease (Carlo & Chatburn, 1988). The mortality rate is about 10%, and RDS is the leading cause of neonatal death and morbidity. Infants who do not require assisted ventilation recover without sequelae, but the clinical course of the very immature infant may be complicated by air leaks in the lungs and BPD. Infants who survive severe RDS often require frequent hospitalization for upper respiratory tract infections and have an increased incidence of neurologic sequelae.

ECMO is a technique of cardiopulmonary bypass modified from techniques developed for open-heart surgery that are used to support heart and lung function (for review of ECMO and implications for pediatric physical therapy, see Caron & Berlandi, 1997, and Pax Lowes & Palisano, 1995). In newborns with acute respiratory failure, the immature lungs are allowed to rest and recover to avoid the damaging effects of artificial ventilation. Because of the need for systemic administration of heparin and the resultant risk of systemic and intracranial hemorrhage, ECMO is reserved for use with infants who are at least 34 weeks of gestational age, weigh more than 2000 g, have no evidence of intracranial bleeding, required less than 10 days of assisted ventilation, and have reversible lung disease (Stork, 1992). ECMO is now used with considerable frequency in support of neonates with severe but reversible respiratory failure, including complicating meconium aspiration, congenital diaphragmatic hernia, sepsis, persistent fetal circulation,

RDS, and BPD (Hibbs, 2001; Martin & Fanaroff, 1992). High-frequency oscillatory ventilation has been used as a bridge from ECMO to conventional ventilation in cases in which conventional ventilation was not initially successful (Schexnayder et al., 1995).

Bronchopulmonary Dysplasia and Chronic Lung Disease of Infancy

Bronchopulmonary dysplasia (BPD) and chronic lung disease of infancy (CLD) are two chronic pulmonary conditions that are caused by incomplete or abnormal repair of lung disease during the neonatal period (Ho, 2002). Infants are diagnosed with BPD when they are 28 days chronologic age and continue to require supplemental oxygen; have an abnormal physical examination with tachypnea, wheezes, and retractions; and have an abnormal chest radiograph (Farrell & Fiascone 1997). CLD is diagnosed at 36 weeks' postmenstrual age, if there is a continued need for supplemental oxygen, abnormal physical examination, and abnormal chest radiograph. The overall incidence varies from 13% to 69% of infants weighing less than 1500 g and requiring ventilation, depending on diagnostic criteria and the neonatal population being studied (Mitchell, 1996). Boys are at greater risk for developing CLDs, perhaps due to a lag of 1 to 2.5 weeks in pulmonary and cerebral maturation in boys (Lauterback et al., 2001).

Pathophysiologic features of BPD include interstitial fibrosis resulting from absorption of intra-alveolar exudate by the alveolar wall during the resolution of RDS. Alveolar collapse may cause parts of the lung to become airless and solid. These nonaerated regions form scars of condensed lung tissue and become fibrotic. Mucosa of the bronchioles becomes dysplastic and inflamed, and there is hypertrophy of the smooth muscle of bronchioles and arterioles. Pulmonary function testing reveals severe maldistribution of ventilation in these infants. They have increased airway resistance, decreased dynamic compliance, and a large increase in the work of breathing.

Prematurity, barotrauma from high pressures used in assisted ventilation, and pulmonary oxygen toxicity are accepted as key causal components of BPD (Bancalari, 1992). Also, the endotracheal tube itself hinders drainage of tracheal secretions and increases both dead space and resistance to airflow. Other factors that may contribute to the pathogenesis of BPD are infection, pulmonary edema resulting from a patent ductus arteriosus or excessive fluid administration, and increased airway resistance. Immaturity of the pulmonary antioxidant systems and neutrophil-generated lung damage are two of the multiple factors associated with the etiology of both BPD and CHD (Ho, 2002; Mitchell, 1996). A genetic disposition to the development has been proposed due to a greater risk of BPD in families with a strong family history of asthma.

Treatment strategies for BPD are aimed at preventing or inhibiting the events that trigger the cascade of pathogenic mechanisms (Goetzman, 1986). These strategies include prevention of premature birth; surfactant replacement therapy; prevention of barotrauma, by using mechanical ventilation for as short a period as is possible, and volutrauma; minimizing oxygen toxicity and increasing antioxidant defenses; control of infection; anti-inflammatory therapy; use of bronchodilators and preventing pulmonary edema by fluid restriction (Ho, 2002). The use of positive end-expiratory pressure during mechanical ventilation, permissive hypercapnia, nasal continuous positive airway pressure, and superoxide dismutase enzymes as an antioxidant are currently under investigation for treatment of BPD and CLD (Ho, 2002).

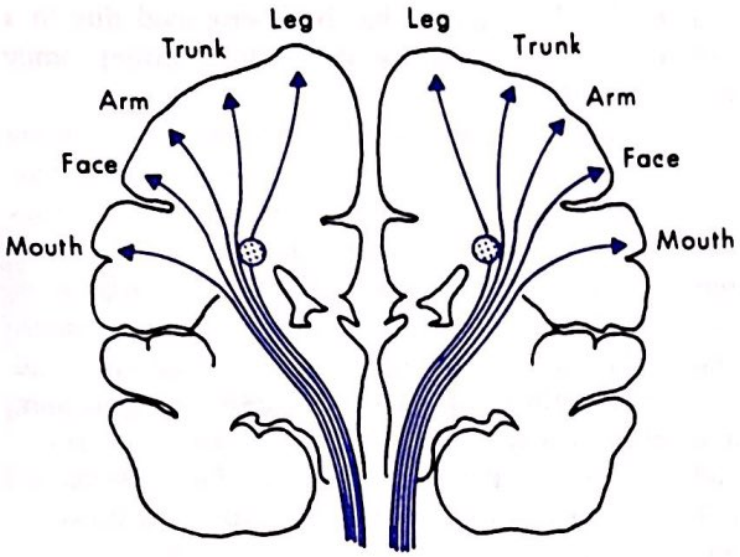
Because infants with BPD are prone to developing cor pulmonale, congestive heart failure, and pulmonary edema, chronic diuretic therapy is instituted. The infant or child with BPD must also be closely monitored for infections, especially those caused by bacteria and fungi. Steroids are used to decrease inflammatory responses and improve lung functions through reduction in pulmonary edema, bronchial edema, and bronchospasm (Kenner, 1998). Other treatments for bronchospasm include inhaled bronchodilator and theophylline therapy (Bancalari, 1992; Davis et al., 1990). Stimulus reduction is also recommended in the nursing care of the infant with BPD (Kenner, 1998).

The incidence of developmental disability, such as mental retardation and cerebral palsy, in infants and children with BPD has been reported to be 29% to 34% (Robertson et al., 1991; Vidyasagar, 1985; Vohr et al., 1991). The important predictors for these disabilities are intracranial hemorrhage and periventricular echodensity, rather than severity of the BPD (Luchi et al., 1991). Transient neuromotor delays have been documented for infants with BPD because of prolonged periods of increased work of breathing, frequent infections, and recurrent hospitalizations (Mitchell, 1996).

Neurologic Complications

Periventricular Leukomalacia

Periventricular leukomalacia (PVL) is a symmetric, nonhemorrhagic, ischemic lesion to the brain of the premature infant (Volpe, 2001). It involves a characteristic distribution of necrosis of white matter dorsal and lateral to the external angles of the lateral ventricles. The area



• **Figure 35-2** Diagram of corticospinal tracts. Dotted circular areas indicate periventricular leukomalacia that would be expected to affect descending fibers for control of the lower extremity. *(From Volpe, JJ. Neurology of the Newborn, 2nd ed. Philadelphia: WB Saunders, 1987, p. 314.)*

affected includes the white matter through which long descending motor tracts travel to the spinal cord from the motor cortex. Because the motor tracts involved in the control of leg movements are closest to the ventricles, and therefore more likely to be damaged, spastic diplegia is the most common clinical sequela (Fig. 35-2). If the lesion extends laterally, the arms may be involved, with resulting spastic quadriplegia. Visual deficits may also result from damage to the optic radiations (Catto-Smith et al., 1985; Papile, 1997).

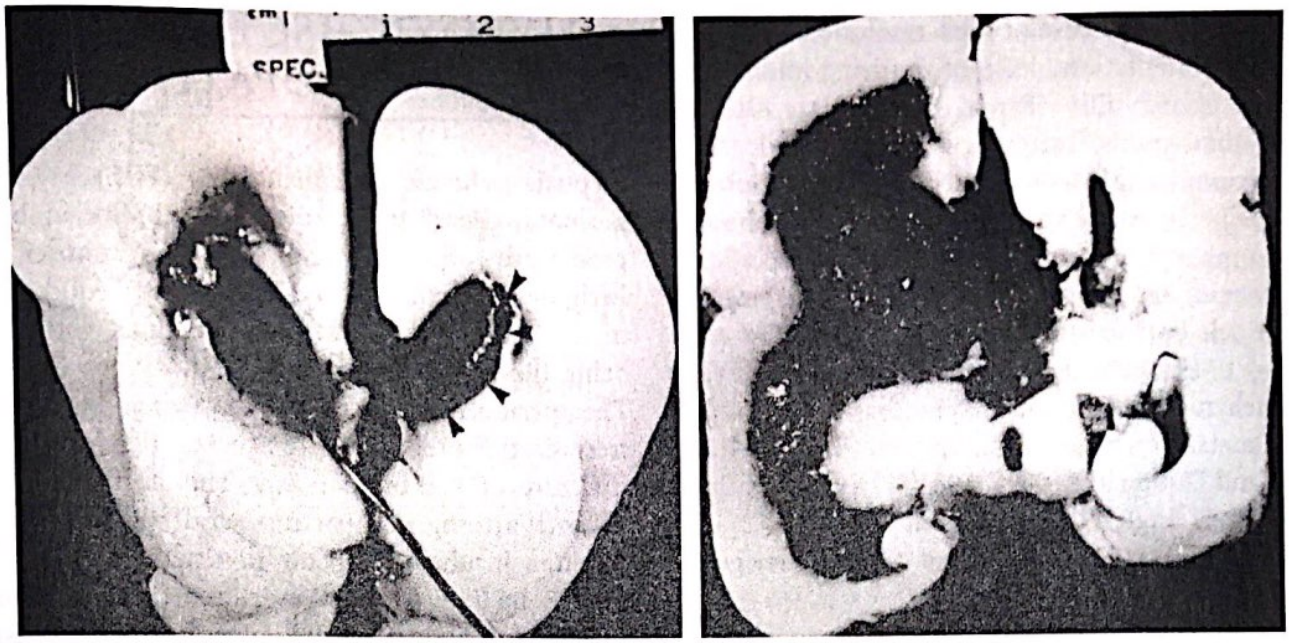
The incidence of white matter damage in premature infants increases with decreased gestational age. The lesion is caused by a reduction in cerebral blood flow in the highly vulnerable periventricular region of the brain where the arterial "end zones" of the middle, posterior, and anterior cerebral arteries meet and is often associated with intraventricular hemorrhage (IVH) (Volpe, 2001). Decreased cerebral blood flow leads to an increase in ischemia and a decrease in antioxidant defenses. The resulting generation of free oxygen radicals and glutamate toxicity are factors that contribute to periventricular leukomalacia. Systemic hypotension associated with a difficult resuscitation at birth and ECMO may be causal factors. Patent ductus arteriosus and severe apneic spells are other contributing factors, particularly after the first week of life (McMenamin et al., 1984; Sweeney & Swanson, 2001; Volpe 2001).

Serial ultrasonography, computed tomography, magnetic resonance imaging, and positron emission tomography are useful diagnostic tools for periventricular leukomalacia (Mizuguchi & Takashima, 2002; Sinha et al., 1990; Volpe 2000). White matter echodensities and echolucencies on high-resolution cranial ultrasonography are predictive of neurologic sequelae associated with cerebral palsy (Leviton & Paneth, 1990; Miller & Murray, 1992). Serial ultrasonographic studies are important because the evolution of periventricular echodensity is related to prognosis. Early periventricular echodensity that resolves during the first weeks of life is not correlated with childhood disability; development of cysts as a result of dissolution of brain tissue secondary to infarction, however, are correlated with cerebral palsy and cognitive deficits (Mantovani & Powers, 1991). Cerebral palsy occurs in more than 90% of infants who develop bilateral cysts larger than 3 mm in diameter in the parietal or occipital areas. Magnetic resonance imaging (MRI) is more sensitive and specific to detection of PVL; however, use with infants who require monitoring equipment is contraindicated (Volpe, 2001).

The basic elements of medical management of PVL include prevention of intrauterine asphyxia, maintenance of adequate ventilation and perfusion, avoidance of systemic hypotension, and control of seizures (Volpe, 2001). Prevention of intrauterine asphyxia includes identification of high-risk pregnancies, fetal monitoring, fetal blood sampling, and cesarean section as indicated. Maintenance of adequate ventilation includes avoiding common causes of hypoxemia such as inappropriate feeding, inserting or removing ventilator connections, painful procedures and examinations, handling, and excessive noise. Adequate perfusion can be maintained with appropriate treatment if the infant exhibits apnea and severe bradycardia. Methods for prevention of systemic hypotension include proper handling, use of volume-expanding medications, treatment of pneumothorax, and abrupt closure of patent ductus arteriosus. Treatment for neonatal seizures includes phenobarbital and calcium-channel blockers, free radical scavengers such as allopurinol, and maternal use of magnesium sulfate as a neuroprotective agent (Volpe, 2001).

Germinal Matrix-Intraventricular Hemorrhage and Periventricular Hemorrhage

Germinal matrix-intraventricular hemorrhage (GM-IVH) is the most common type of neonatal intracranial hemorrhage and is characteristic of the premature infant of less than 32 weeks of gestational age and weighing less than 1500 g. GM-IVH typically occurs in preterm infants with RDS requiring ventilation. An inverse relationship exists between gestational age and incidence of GM-IVH. The average incidence has been reported at 15% in preterm infants who are in the aforementioned categories



• **Figure 35-3** Coronal section of the cerebrum showing intraventricular hemorrhage. *(From Volpe, JJ. Neurology of the Newborn, 2nd ed. Philadelphia: WB Saunders, 1987, p. 314.)*

for gestational age and weight (Volpe, 2001). Most hemorrhages occur within the first 2 postnatal days, and 90% occur within 72 hours of birth (Papile, 1997; Volpe, 2001). This lesion involves bleeding into the subependymal germinal matrix, which is a gelatinous area that contains a rich vascular network supplied mainly by Heubner's artery, a branch of the anterior cerebral artery; branches of the middle cerebral artery, and internal carotid. This matrix is prominent from 26 to 34 weeks of gestation and is usually gone by term. The vessels that traverse the matrix are primitive in appearance, with a single layer of endothelium without smooth muscle, elastin, or collagen, and the area is devoid of supportive stroma. The site of origin of hemorrhage is from these primitive capillaries. In a small number of preterm infants, hemorrhage may occur from the choroid plexus or roof of the fourth ventricle (Fig. 35-3). GM-IVH pathogenesis includes fluctuating cerebral blood flow, a decrease followed by an increase in cerebral blood flow, an increase in cerebral venous pressure, and platelet and coagulation disturbance (Volpe, 2001).

Neuropathologic complications of IVH are hydrocephalus, germinal matrix destruction, cyst formation, and accompanying hypoxic-ischemic lesions. A four-level grading scale based on ultrasound scan has been developed by Papile to classify hemorrhages (Papile et al., 1983). Grade I is an isolated germinal matrix hemorrhage. Grade II is an IVH with normal-sized ventricles that occurs when hemorrhage in the subependymal germinal matrix ruptures through the ependyma into the lateral ventricles. Grade III includes an IVH with acute ventricular dilation, and grade IV involves a hemorrhage into the periventricular white matter.

It has recently been shown that the cause of periventricular hemorrhage is not a simple extension of blood into the cerebral white matter from the germinal matrix, but that the lesion is a hemorrhagic venous infarction with the major cause being obstruction of blood flow in the terminal vein caused by GM-IVH (Volpe, 2001). Periventricular hemorrhagic infarction is most often unilateral or markedly asymmetric, and grossly hemorrhagic. Accompanying lesions include periventricular leukomalacia, pontine neuronal necrosis, and hydrocephalus. Catastrophic IVH is the least common and occurs in infants with severe hemorrhaging. Volpe (2001) describes three IVH syndromes: catastrophic, saltatory, and clinically silent. Clinical correlates for the catastrophic IVH include coma, respiratory abnormalities, generalized tonic seizures, decerebrate posturing, eyes fixed to vestibular stimulation, and flaccid quadriparesis. Presenting signs of the saltatory syndrome include an alteration in level of consciousness, decrease in quantity and quality of spontaneous and elicited movement, hypotonia, and eye abnormalities such as downward vertical drift. These neurologic signs are subtle and often difficult to detect in early weeks of life via clinical examination. A more sensitive finding is an unexplained fall in hematocrit.

Serial portable cranial sonography is the procedure of choice in the diagnosis, identification, grading, and

timing of GM-IVH because of high-resolution imaging, portable instrumentation, lack of ionizing radiation, and relative affordability (Papile, 1997; Volpe, 2001). Computed tomography scanning is useful for identification of complicating lesions such as posterior fossa lesions, subdural hemorrhage, and parenchymal abnormalities. Lumbar puncture findings for IVH include a high number of red blood cells and increased protein content, which both correlate with the severity of the hemorrhage. MRI provides excellent imaging of IVH and parenchymal details but requires transport, and absence of metallic monitoring equipment (Volpe, 2001). Dubowitz and Dubowitz (1981a, 1981b) have noted that a tight popliteal angle (130° or less in infants up to 31 weeks of gestation and 110° or less at 32 to 35 weeks of gestation) is a sign of IVH.

Methods to prevent IVH include measures to prevent premature birth, transport in utero and prenatal administration of corticosteroid to women at risk for preterm birth (Volpe, 2001; Crowley, 2002). Treatment includes support of cerebral perfusion by maintaining arterial blood pressure and avoidance of cerebral hemodynamic disturbances caused by rapid volume expansion, pneumothorax, increased arterial blood pressure, and hypoxemia. Serial ultrasound scans to monitor ventricular size and ventriculostomy or shunting for hydrocephalus are also important treatment components (Volpe, 2001). Prophylactic measures include transport of the infant in utero to a level III nursery, limiting handling, optimal management of labor and delivery, cesarean section before active phase of labor, and medications such as phenobarbital, indomethacin, and ethamsylate (to reduce capillary bleeding). Vitamins E and K are currently under investigation to reduce capillary bleeding. When hemorrhage has occurred, medical management includes maintenance of cerebral perfusion through control of blood pressure and decrease in intracranial pressure through lumbar puncture and ventricular drainage.

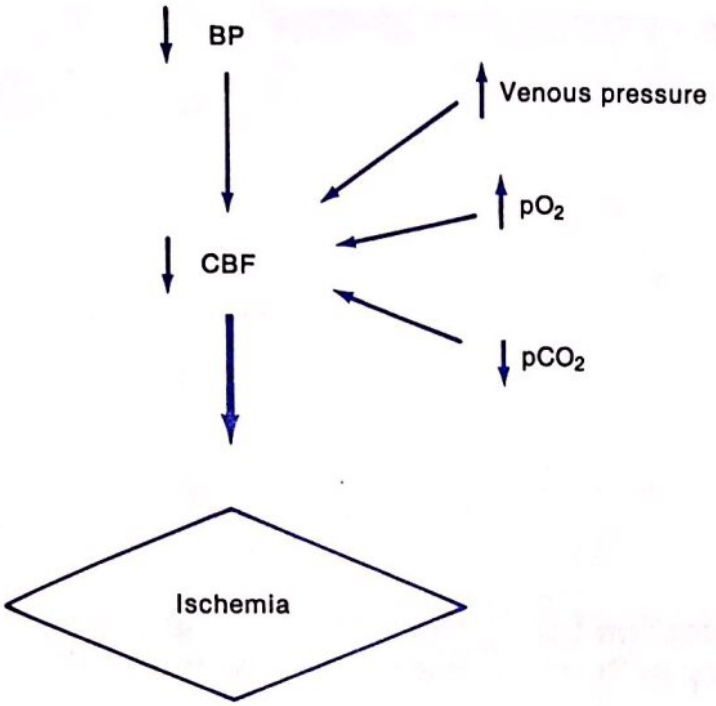
Short-term outcome of the infant with GM-IVH is correlated with the severity of the hemorrhage, but long-term outcome depends on the degree of associated parenchymal injury (Volpe, 2001). Volpe reported that for neonates with parenchymal injury greater than 1 cm, the mortality rate was 59% and the rate of major neurologic sequelae such as spastic hemiparesis or asymmetric spastic quadriplegia was 87%. In addition, 73% of the survivors had cognitive impairments. The risk of major neurologic disability in LBW preterm infants increases with each higher grade of IVH (McMenamin et al., 1984). The incidences of neurologic sequelae for grades I, II, and III are 5%, 15%, and 35%, respectively (Volpe, 2001).

HYPOXIC-ISCHEMIC ENCEPHALOPATHY

Hypoxic-ischemic encephalopathy (HIE) is the major perinatal cause of neurologic morbidity in both the premature and full-term infant (Vannucci, 1997). Perinatal asphyxia results in hypoxemia (reduced amount of oxygen in the blood) and ischemia, with ischemia being the most important form of oxygen deprivation. The period of reperfusion is when many of the complications that affect the metabolism, function, and structure of the brain occur. The relative importance of antepartum, intrapartum, and postnatal hypoxic-ischemic insults is difficult to determine. Major signs of HIE include seizures and abnormalities in state of consciousness, tone, posture, reflexes (especially disturbed suck, swallow, gag, and tongue movements), respiratory pattern, and autonomic function. Conditions associated with increased risk for fetal asphyxia are altered placental exchange, reduced maternal blood flow to the placenta, and decreased maternal oxygen saturation. Altered placental exchange may result from abruption, placenta previa, postmaturity, prolapsed umbilical cord, umbilical cord around the neck, and placental insufficiency. Maternal hypotension may reduce the blood flow to the placenta, and maternal hypoventilation, hypoxia, or cardiopulmonary disease may decrease maternal oxygenation. Intrapartum insults include those related to traumatic delivery, prolonged labor, and acute placental or cord problems. Prolonged partial asphyxia from any cause sets into motion a spiral of cytotoxic edema, with impaired cerebral blood flow, which leads to cerebral necrosis or ulegyria (Fig. 35-4). Infants with HIE commonly have disturbances of pulmonary, cardiovascular, hepatic, and renal functions.

Therapeutic care of an infant at risk for development of HIE includes careful monitoring by serial neurologic assessments, detection of reduced cerebral perfusion pressure, and assessment of the structural status of brain with computed tomography, ultrasound, magnetic resonance imaging, magnetic resonance spectroscopy, technetium scan, and electroencephalography or evoked potential tests (Volpe, 2001). Treatment principles include prevention of intrauterine asphyxia; maintenance of adequate ventilation, perfusion, and blood glucose levels; and control of seizures and brain swelling.

Preventive measures for intrauterine asphyxia include antepartum assessment and identification of the high-risk pregnancy, electronic fetal monitoring, fetal blood sampling, and cesarean section when necessary. Maintenance of adequate oxygenation is essential to prevent



• **Figure 35-4** Model for production of cerebral edema. Decreased blood pressure (BP) leads to decreased cerebral blood flow (CBF) and ischemia. *(From Campbell, SK: Clinical decision making: Management of the neonate with movement dysfunction. In Wolf, SL [Ed.]. Clinical Decision Making in Physical Therapy. Philadelphia: FA Davis, 1985, p. 299.)*

additional injury. Volume expansion with the use of drugs to increase blood pressure (pressor agents) is used to maintain cerebral blood flow. Partial exchange transfusion may be done if the infant's hematocrit is low to decrease hyperviscosity and enhance tissue oxygenation. Sodium bicarbonate and glucose are administered to treat severe, persistent metabolic acidosis and hypoglycemia, respectively. The use of barbiturates is being studied because they reduce the energy requirements of the brain and have been shown to decrease intracranial pressure in adults. Phenobarbital is the drug of choice for seizure control, with lorazepam as adjunctive therapy.

Sarnat and Sarnat (1976) identified three stages of encephalopathy after asphyxia. Stage 1 lasts less than 24 hours and is characterized by hyperalertness, low-threshold Moro and stretch reflexes, and normal muscle tone and electroencephalogram. The duration for stage 2 is approximately 5 days; clinical signs include lethargy, mild hypotonia, strong distal flexion, hyperactive stretch reflexes, weak suck and Moro reflexes, strong tonic neck reflex, and multifocal seizures. Stage 3 is stupor, which lasts from a few hours to 4 weeks, with flaccidity alternating with decerebrate posturing, absent reflexes, response only to strong noxious stimuli, occasional seizures, and an isopotential electroencephalogram.

When the infant with hypoxic-ischemic insult has a normal neurologic examination by 1 week of age, sequelae are minimal. Those who continue to have an abnormal neurologic examination by 3 weeks of age, however, are at risk of developing major neurologic sequelae (Harper & Yoon, 1987). The neurologic sequelae of HIE include motor deficits with or without mental retardation, seizures, or both. Neuropathologic classifications of HIE include selective neuronal necrosis, periventricular leukomalacia, parasagittal cerebral injury, and focal ischemic brain necrosis (Volpe, 2001).

SELECTIVE NEURONAL NECROSIS

Selective neuronal necrosis, the most common type of HIE, is the death of neurons in a widespread but characteristic pattern and commonly accompanies the other manifestations of HIE. The major sites of neuronal necrosis include the hippocampus of the cerebral cortex and parts of the diencephalon, basal ganglia, pons, medulla, cerebellum, thalamus, brainstem, and spinal cord (Cabanas et al., 1991) (Box 35-2, Table 35-1, and Fig. 35-5). Topography is related to the severity and duration of the HIE and the gestational age of the infant or fetus (Volpe, 2001). The pathogenesis of selective neuronal necrosis includes regional vascular factors because neuronal injury is more marked in vascular border zones, regional metabolic factors, and the regional distribution of glutamate receptors, particularly the NMDA type (Volpe, 2001). Clinical findings include stupor, coma, seizures, hypotonia, and problems in ocular, sucking, and tongue movements. Long-term sequelae include cognitive impairment, spastic quadriparesis, seizure disorder, ataxia, bulbar and pseudobulbar palsy, hyperactivity, impaired attention, and atonic quadriparesis.

Status marmoratus is neuronal loss, gliosis, and hypermyelinization of the basal ganglia and thalamus. Impairments are not fully manifested until the latter part of the first year of life, although injury occurs in the perinatal period (Volpe, 2001). The pathogenesis of this lesion is related primarily to glutamate-induced neuronal death, as well as regional circulatory and metabolic factors (Volpe, 2001). Neonatal findings are unknown at this time, but long-term clinical sequelae include choreoathetosis, cognitive impairment, and spastic quadriplegia.

PARASAGITTAL CEREBRAL INJURY

Parasagittal cerebral injury results in a lesion of the cerebral cortex and subcortical white matter. This injury is usually bilateral and symmetric, with the parietal-occipital regions most affected (Volpe, 2001). The areas of

Box 35-2 Selective Neuronal Necrosis in Neonatal Hypoxic-Ischemic Encephalopathy—Major Sites

Cerebral cortex —hippocampus, supralimbic cortex
Diencephalon —thalamus, hypothalamus, and lateral geniculate body
Basal ganglia —caudate, putamen, and globus pallidus
Midbrain —inferior colliculus, oculomotor and trochlear nuclei, red nucleus, substantia nigra, and reticular formation
Pons —motor nuclei of trigeminal and facial nerves; dorsal, ventral cochlear nuclei; reticular formation; and pontine nuclei
Medulla —dorsal motor nucleus of vagus nerve, nucleus ambiguus, inferior olivary nuclei, cuneate and gracilis nuclei
Cerebellum —Purkinje cells, dentate, and other roof nuclei

From Volpe, JJ. Neurology of the Newborn, 2nd ed. Philadelphia: WB Saunders, 1987, p. 213.

TABLE 35-1 Sites of Particular Predilection for Apparent Hypoxic-Ischemic Selective Neuronal Injury in Premature and Term Newborns*

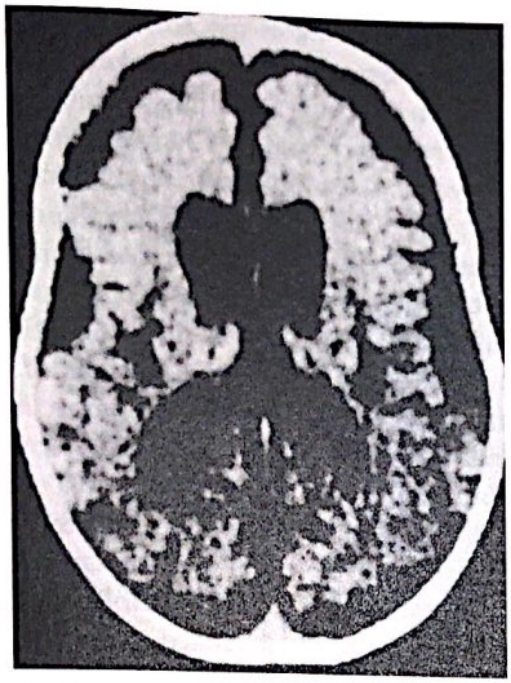
BRAIN REGION	PREMATURE	TERM
Cerebral neocortex		+†
Hippocampus		
Sommer's sector		+
Subiculum	+	
Basal ganglia/thalamus	=†	=
Brainstem		
Cranial nerve nuclei	=	=
Pons (ventral)	+	
Inferior olivary nuclei	+	
Cerebellum		
Purkinje cells		+
Internal granule cells	+	
Spinal cord		
Anterior horn cells (alone)		+
Infarction (including anterior horn cells)	+	

*See text for references.
†+, Relatively more common in term versus premature newborn (or vice versa); =, equally common in term and premature newborns.

necrosis are the border zones between the end fields of the major cerebral arteries. Clinical signs in the neonatal period include proximal limb weakness, especially in the upper extremities. Long-term sequelae include spastic quadriparesis and specific intellectual deficits such as delay in development of language, visuospatial abilities, or both.

FOCAL ISCHEMIC BRAIN NECROSIS AND CAVITATION

The category of focal ischemic brain necrosis includes large, localized areas of neuronal death in the distribution of single or multiple major blood vessels in the cerebral cortex and subcortical white matter. Most of these lesions are unilateral and involve the middle cerebral artery. The focal ischemic brain necrosis occurs perinatally as the result of cerebrovascular insufficiency secondary to malformation of vessels, arterial obstruction due to thrombi or emboli, or systemic circulatory insufficiency near the end of the second trimester. Resolution of neuronal necrosis results in the formation of cavities. Neurologic features during the neonatal period include hemiparesis, quadriparesis, and stereotyped and non-habituating reflex responses. This lesion results in spastic



• **Figure 35-5** Computed tomography scan of selective cortical neuronal necrosis of a 4-week-old infant who experienced severe perinatal asphyxia. Note cortical atrophy and white matter injury. *(From Volpe, JJ. Neurology of the Newborn, 2nd ed. Philadelphia: WB Saunders, 1987, p. 245.)*

hemiparesis in the case of a limited focal lesion, mental retardation, and seizure disorder (Volpe, 2001).

NECROTIZING ENTEROCOLITIS

Necrotizing enterocolitis (NEC) is an acute inflammatory disease of the bowel that occurs most frequently during the first 6 weeks of life in premature infants weighing less than 2000 g (Whaley & Wong, 1991). Although the precise cause of the disease is not known, several factors appear to play a role in the pathogenesis of NEC. These factors include intestinal ischemia, infectious agents and toxins, and enteral formula alimentation (McElhinney et al., 2000; McGuire & Antony, 2003). Diminished blood supply results in death of mucosal cells lining the bowel wall, decreasing secretion of lubricating mucus. The thin bowel wall becomes susceptible to proteolytic enzymes, swells, breaks down and is permeable to exotoxins. Gas-forming bacteria invade the damaged area to produce pneumatosis intestinalis, air in the submucosal or subserosal surfaces of the bowel (Hockenberry et al., 2003).

Diagnosis is made by physical examination, laboratory studies, and radiography. Vomiting, abdominal distention, bloody stools, retention of stools, lethargy, decreased urine output, and alterations in respiratory status are signs of NEC (Dolgin et al., 1998; Hockenberry et al., 2003). Medical care of infants with NEC includes parenteral alimentation, gastric suction, and administration of broad-spectrum antibiotics (Cressinger et al., 1992). Laboratory findings include anemia, leukopenia, and electrolyte imbalance. Breath hydrogen measurements have been found to be 99% effective in detecting absence of the disease and are suggested as an aid to diagnosis of NEC (Cheu et al., 1989). Abdominal radiographs are performed every 6 to 8 hours to detect progressive intestinal obstruction or possible perforation.

Preventive treatment includes withholding oral feedings for at least 24 to 48 hours from infants who suffered birth asphyxia. Breast milk is the preferred enteral nutrient because it allows for passive immunity (Hockenberry et al., 2003). Kamitsuka and associates (2000) reported that implementation of a standard feeding protocol in infants weighing 1250 to 2500 g and who are less than 35 weeks' gestation reduced the occurrence of NEC by 24%. Medical treatment of confirmed NEC includes discontinuation of all oral feedings, abdominal decompression via nasogastric suction, administration of intravenous antibiotics, and correction of fluid and electrolyte imbalances (Hockenberry et al., 2003). Surgical intervention is indicated when there is radiographic evidence of fixed, dilated intestinal loops accompanied by intestinal distention (Cressinger et al., 1992). Surgical procedures include intestinal decompression by nasogastric tube placement or gastrostomy, resection of necrotic bowel, and diversion of the proximal fecal stream by ileostomy, jejunostomy, or colostomy. Although enterostomy is a lifesaving procedure, it has been reported to be a major cause of morbidity (O'Connor & Swain, 1998).

NEC is the most common cause of death in neonates undergoing surgery and is the most frequent and lethal gastrointestinal disease of premature infants (Nadler et al., 2001). Factors that are predictive of poor outcome include prior enteral feeding, PDA, indomethacin use, and perforation (Wang et al., 2002). Mortality rates in the 1990s (9% to 28%) are considerably lower that the rates reported in the 1960s and 1970s (24% to 65%) (Kenner & Lott, 2003). The average mortality rate after surgery is 30% to 40%. Stricture formation, which is abnormal narrowing of the intestines, occurs in 25% to 35% of the survivors of medical or surgical treatment and causes failure to thrive, feeding abnormalities, diarrhea, or bowel obstruction (Cressinger et al., 1992).

RETINOPATHY OF PREMATURITY

Retinopathy of prematurity (ROP) is a lesion caused by an ischemic event that interferes with the development of retinal blood vessels. Cessation of this normal retinal

angiogenesis is followed by hyperpoliferative neovascular response to retinal ischemia. Subsequently, there is vascular proliferation of retinal capillaries into the hypoxic area. New vessels proliferate toward the lens, and the aqueous humor and then the vitreous humor become turbid. The retina becomes edematous, and hemorrhages separate the retina from its attachment (Phelps, 1992). The outcome of the disease ranges from normal vision to total loss of vision if there is advanced scarring from the retina to the lens resulting in retinal detachment (Whaley & Wong, 1991). ROP was called retrolental fibroplasia in the early 1940s and was virtually eliminated with the severe restriction of oxygen use between 1950 and 1970. The condition has recurred as one of the major causes of disability in preterm infants as a result of the increased survival of VLBW infants. The incidence of ROP increases with lower gestational age, lower birth weight, and BPD (Holmstrom et al., 1998).

The pathogenesis of ROP is abnormal angiogenesis (development of the blood vessels) in the retina. Vascular endothelial growth factor (VEGF) seems to play a key role in the normal and abnormal angiogenesis. The VEGF gene is responsive to oxygen tension. Hypoxia stimulates VEGF transcription and hyperoxia decreases VEGF transcription. When oxygen is administered to premature infants, relatively hyperoxic conditions occur and VEGF levels decrease. Supplemental oxygen supports the avascular retina, but as oxygen is weaned the retina becomes ischemic. This ischemia stimulates VEGF transcription and angiogenesis resumes often in an uncontrolled hyperproliferative manner (Holstrom et al., 1998).

The severity of ROP is classified in five stages (Stout & Stout, 2003). In stage 1 there is a visible line of demarcation between posterior vascularized retina and the anterior avascular retina. In stage 2 pathologic neovascularization is confined to the retina that appears as a ridge at the vascular/avascular junction. Stage 3 includes new vascularization and migration into the vitreous gel. Stage 4 is defined by a subtotal retinal detachment. Stage 5 is complete retinal detachment.

Hyperoxia, shock, asphyxia, hypothermia, vitamin E deficiency, and light exposure have been implicated as possible pathogenic factors (Kretzer & Hittner, 1986). Antenatal dexamethasone administration appears to be associated with a decreased incidence of development of ROP of stage 2 or higher (Higgins et al., 1998). Light reduction was not shown to be effective in altering the incidence of ROP (Reynolds et al., 1998).

Prevention and treatment include oxygen administration at Pao_2 between 50 and 70 mm Hg and administration of vitamin A (which is still under investigation) (Darlow & Graham, 2002). All premature infants given supplemental oxygen are at risk for ROP and should be screened. Guidelines approved by the American Academy of Pediatrics include two screening examinations 4 to 6 weeks after birth or within 31 to 33 weeks postconceptual age, whichever is later. Subsequent intervals for examination are based on initial findings (Stout & Stout, 2003). A study conducted in Sweden recommended that screening criterion be lowered to 31 weeks or less to identify infants with severe ROP (Larsson & Holmstrom, 2002). Surgical intervention can be divided into two overlapping objectives: treatment of neovascular process with retinal cryotherapy and surgical intervention for retinal detachment (laser photocoagulation, cryotherapy, vitrectomy, and scleral buckling) (Stout & Stout 2003; Kretzer & Hittner, 1986). Supplemental oxygen with target oxygen saturation of 99% with Po_2 of no higher than 100 mm Hg was associated with regression of prethreshold ROP, without appearing to arrest retinal vascular maturation (Gaynon et al., 1997). Implementation of an oxygen management policy that included strict guidelines for increasing and weaning of FIo_2 (fraction of inspired oxygen), monitoring of oxygen saturation parameters in delivery room, in-house transport, and hospitalization for infants 500 to 1500 g birth weight resulted in a decreased incidence of ROP stages 3 to 4 and decreased the need for laser treatment (Chow et al., 2003). Surgical outcome ranges from complete recovery or mild myopia to blindness, depending on the extent of the disease.

HYPERBILIRUBINEMIA

Hyperbilirubinemia, or physiologic jaundice, is the accumulation of excessive amounts of bilirubin in the blood. Bilirubin is one of the breakdown products of hemoglobin from red blood cells. This condition is seen commonly in premature infants who have immature hepatic functions, an increased hemolysis of red blood cells as a result of high concentrations of circulating red blood cells, a shorter life span of red blood cells, and possible polycythemia from birth injuries (Hockenberry et al., 2003). The primary goal in treatment of hyperbilirubinemia is the prevention of kernicterus, which is the deposition of unconjugated bilirubin in the brain, especially in the basal ganglia and hippocampus. LBW infants of less than 2000 g receive phototherapy at 24 hours of life for 96 hours, regardless of bilirubin concentration. Phototherapy is administered by placing 8 to 10 fluorescent lamps 12 to 16 inches above the infant (Gartner & Lee, 1992). The infant is positioned in an open radiant warmer or incubator and the eyes are shielded to avoid retinal damage. Studies show that on/off cycles of

more than 1 hour are as effective as continuous treatment. Infants who received traditional phototherapy for 23 hours and kangaroo care (skin to skin contact with parent) with fiberoptic panel held against their back showed comparable declines in bilirubin levels (Ludington-Hoe & Swinth, 2001). A technique of fiberoptic phototherapy uses light from a halogen lamp transmitted through a fiberoptic bundle to a blanket that is wrapped around the infant (Biliblanket) (Gartner & Lee, 1992).

If phototherapy is not effective in reducing the total serum bilirubin concentrations to acceptable levels, or if there is a rapidly rising bilirubin level, exchange transfusion is done (Shaw, 1998). In this technique, approximately 85% of the infant's red blood cells are replaced. Care must be taken so as not to disrupt cerebral blood flow and intracranial pressure (Volpe, 2001). Initial human trials have shown beneficial effects of metalloporphyrins such as tin (Sn) mesoporphyrin and tin protoporphyrin used as prophylaxis and treatment to reduce hyperbilirubinemia. These substances are inhibitors of heme oxygenase, which is an enzyme in the synthesis of bilirubin that limits the rate of degradation of heme to bile (Gartner & Lee, 1992; Kenner & Lott, 2003). Infants with severe hyperbilirubinemia unresponsive to phototherapy whose parents are Jehovah's Witnesses and rejected exchange transfusion have been successfully treated with Sn-mesoporphyrin (Kappas et al., 2001). Less frequently used therapy is enterohepatic circulation enhancement by administration of agar that binds to bilirubin in the intestine, inhibits resorption, and promotes excretion. Conjugation of bilirubin has been induced as a side effect of phenobarbital (Volpe, 2001).

NEONATAL SEIZURES

Seizures are the most frequent and distinct neurologic signs that occur in the neonatal period (Volpe, 2001; Brann & Wiznitzer, 1992). Seizures are the most frequent overt sign of neurologic disorders (Volpe, 2001). HIE, low birth weight, low gestational age, jaundice during the last 3 days, maternal disease in the last 2 years before pregnancy, intrapartum fever, jaundice during the first 3 days of life, and the need for cardiopulmonary resuscitation are related to neonatal seizures (Arpino et al., 2001; Lieberman et al., 2000). Most seizures occur between the second and fifth days of life, and 85% occur in the first 15 days of life with a sharply increased incidence in infants less than 1500 g. A seizure during the neonatal period is a medical emergency, because it may indicate a life-threatening disease or disorder that can produce immediate and irreversible brain damage. Repeated neonatal seizures may also result in decreased DNA content and brain cell number. The causes of neonatal seizures include hypoxic-ischemic encephalopathy secondary to perinatal asphyxia, intracranial hemorrhage, hypoglycemia, hypocalcemia, intracranial infection, developmental defects, and drug withdrawal (Volpe, 2001).

Clinical manifestations of seizures in neonates differ greatly from those in older infants and children because of the immaturity of the brain. Clinical signs in the neonate include facial, oral, lingual, and ocular movements and autonomic manifestations such as apnea and changes in blood pressure, heart rate, and pupil size (Box 35-3). Abnormal movements and alteration of tone in the trunk and extremities, including rowing and bicycling movements, are also clinical signs of neonatal seizure (Mizrahi & Kellaway, 1987). Symptoms of seizures in premature infants include staring, nystagmus, apnea, hiccough, and chewing, ocular, and pedaling movements (Volpe, 2001). Treatment includes administration of anticonvulsants such as phenobarbital, phenytoin, and diazepam to control seizures and intravenous glucose for hypoglycemia.

Approximately 25% to 35% of all infants with neonatal seizures later exhibit cognitive impairment, motor impairment, or both. Seizures that occur in conjunction with perinatal asphyxia, severe IVH, intracranial infection, and prematurity with prolonged hypoglycemia have poor prognoses, with possible permanent neurologic sequelae

Box 35-3 Characteristics of Subtle Seizures in Premature and Full-Term Infants

- Ocular-tonic horizontal deviation of eyes (jerking) and sustained eye opening with ocular fixation
- Eyelid blinking or fluttering
- Sucking, smacking, drooling, and other oral-buccal-lingual movements
- "Swimming," "rowing," and "pedaling" movements
- Apneic spell

From Volpe, JJ. Neurology of the Newborn, 2nd ed. Philadelphia: WB Saunders, 1987, p. 134.

such as cerebral palsy or cognitive impairment. Seizures that occur with subarachnoid hemorrhage without asphyxia or as a result of metabolic disorders have good prognoses if treatment begins early (Volpe, 2001).

FETAL ALCOHOL SYNDROME

Chronic alcohol exposure in utero may result in a multitude of symptoms at birth, including withdrawal symptoms of irritability, tremors, apnea, and seizures (Volpe, 2001; West et al., 1998). When severe, phenobarbital or paregoric (morphine) may be used to control withdrawal symptoms (Harper & Yoon, 1987). Alcohol rapidly crosses the placenta and the blood-brain barrier of the fetus, and there is a dose-dependent relationship between maternal alcohol intake in the first weeks of pregnancy and the occurrence of features of the fetal alcohol syndrome (FAS).

The full manifestation of FAS, one of the most common causes of mental retardation in the world, is characterized by a triad of symptoms composed of growth deficiency; cardiac defects; and CNS disturbance, such as microcephaly, mental retardation, and dysmorphology (including facial, genital, and joint abnormalities) (Barbour, 1990; Volpe, 2001). The severity of cognitive disability is correlated with the severity of dysmorphic features. Prenatal exposure to alcohol causes deficits in all domains of adaptive functioning including problems with socialization behavior of young children (Whaley et al., 2001).

In utero alcohol exposure is related to a wide continuum of effects ranging from full FAS to partial FAS, also referred to as fetal alcohol effects, to more subtle effects, such as neurologic disorders without dysmorphology (Scott et al., 1991). These subtle neurologic effects include hyperactivity, delayed language development, fine and gross motor problems, slowed reaction time, and problems with judgment and comprehension (Autti-Ramo & Granström, 1991; Scott et al., 1991). The Institute of Medicine has recently proposed the terms alcohol-related neurodevelopmental disorder (ARND) and alcohol-related birth defects (ARBD) to replace FAE (fetal alcohol effects) and FAS in describing conditions in which there is a history of maternal alcohol exposure and an outcome that confirms effects of such exposure (American Academy of Pediatrics, 2000). Long-term effects of prenatal alcohol exposure include deficits in several areas of intellectual functioning, including information processing, short-term memory, encoding, and preacademic skills (Coles et al., 1991; Little et al., 1989). Streissguth (1986) reported that 24% of children born to binge drinkers and 15% born to nonbinge drinkers were receiving special education services at 7.5 years of age. In addition to cognitive deficits, behavioral and communication problems lead to maladaptive social functioning, including impulsive behavior, anxiety, and dysphoria (Volpe, 2001).

NEONATAL ABSTINENCE SYNDROME

Maternal use of narcotics during pregnancy leads to the development of dependency on that drug by the fetus. The most commonly used drugs are heroin, methadone, and cocaine, and there is often maternal use of several drugs during pregnancy (Finnegan, 1988; Kenner & Lott, 2003). Women who use cocaine during pregnancy receive less prenatal care and are more likely to smoke, consume alcohol, be malnourished, and be exposed to sexually transmitted diseases (Tronick & Beeghly, 1999). The fetus experiences withdrawal or neonatal abstinence syndrome (NAS) when the mother is withdrawn from her drug or drugs or when the fetus is delivered (D'Apolito & Hepworth, 2001). The onset of withdrawal symptoms usually occurs within the first 72 hours after birth. (D'Apolito & Hepworth, 2001). Symptoms of withdrawal include neurologic, gastrointestinal, and respiratory signs as well as autonomic dysfunction (Kenner & Lott, 2003). Neurologic symptoms include signs of CNS excitability such as hyperactivity, irritability, tremors, seizures, apnea, increased muscle tone, inability to sleep, hyperactive deep tendon reflexes, and poor coordination. Gastrointestinal symptoms include hyperactive sucking soon after birth and then disorganized, ineffective sucking of reduced rate and pressure that is poorly coordinated with swallowing. Respiratory signs in NAS include tachypnea, irregular respirations, rhinorrhea, stuffy nose, nasal flaring, chest retractions, intermittent cyanosis, and apnea. Regurgitation and/or aspiration may occur. Signs of autonomic dysfunction include frequent sneezing, yawning, mottling of the skin, excessive tearing and generalized sweating, and shrill crying (Dixon, 1989; Hayford et al., 1988; Volpe, 2001, Kenner & Lott, 2003).

Premature delivery, meconium aspiration, low birth weight, small for gestational age, decreased head circumference, and abruptio placentae are common complications of labor and delivery (Chasnoff, 1988; Handler et al., 1991; Kenner & Lott, 2003). Disturbances in the behavioral organization and interactive abilities of these neonates make early bonding and attachment difficult (Hume et al., 1989). Follow-up studies indicate growth retardation, long-term intellectual impairment, and

learning difficulties such as problems with sustained attention and with processing in the visual modality in children exposed to drugs in utero (Bauman & Levine, 1986; Coles, et al., 2002) and problems in adaptive and social functioning (Whaley et al., 2001; Free et al., 1990; Hume et al., 1989; Lewis et al., 1989; Davis & Templer, 1988). Treatment of NAS includes pharmacologic therapy for narcotic withdrawal, but infants exposed to only cocaine do not usually require such treatment (Askin & Diehl-Jones, 2001). Reducing environmental stimulation and using techniques such as swaddling and flexed positioning are effective strategies for calming infants experiencing CNS irritation (Askin & Diehl-Jones, 2001). Small, frequent feedings and additional calories may be needed.

EXPOSURE TO HUMAN IMMUNODEFICIENCY VIRUS

The risk of infants born to women with a positive HIV titer developing HIV infection was estimated to be 10% to 40% in 1995 (Volpe, 2001). Wortley and associates (2001) reported that with good prenatal surveillance for HIV with use of ZDV, only 8% of children exposed to HIV in utero were infected. Isolation is not necessary for the neonate exposed to HIV. An infant born to an HIV-positive mother should not be breastfed, as breast milk is a vehicle for transmission of the virus between mother and infant. Using virologic techniques such as HIV culture, polymerase chain reaction, and immune complex-dissociation, diagnosis of HIV infection can be made in almost 50% of infants at birth and 95% or more of infants by 1 to 3 months of age. Infection early in fetal development may lead to microcephaly, cerebral atrophy, basal ganglia and white matter calcification, calcific vasculopathy of the CNS, spinal cord myelin loss, and facial anomalies (Volpe, 2001). Infants exposed to HIV should be monitored for signs of infection, such as failure to thrive, weight loss, temperature instability, and diarrhea, and assessed for opportunistic infections, such as herpes simplex virus infection, cytomegalovirus infection, lymphoid interstitial pneumonia, and viral, fungal, or protozoal infections. Breastfeeding should not occur because of transmission of the virus and cytomegalovirus (Pursell, 1998; Mussi-Pinhata et al., 1998). Because neonates with HIV infection are usually asymptomatic, they are not admitted to the NICU unless there are problems unrelated to HIV. It should be noted, however, that infants who were seropositive exhibited significantly lower scores on the Bayley Scales of Infant Development than infants who were seronegative (Aylward et al., 1992).

BIRTH INJURIES RELATED TO THE NERVOUS SYSTEM

Birth injuries are those sustained during labor and delivery and represent an important cause of neonatal morbidity (Mangurten, 1992). The most common traumatic lesions related to the nervous system include caput succedaneum, linear skull fracture, and brachial plexus palsy.

CAPUT SUCCEDANEUM

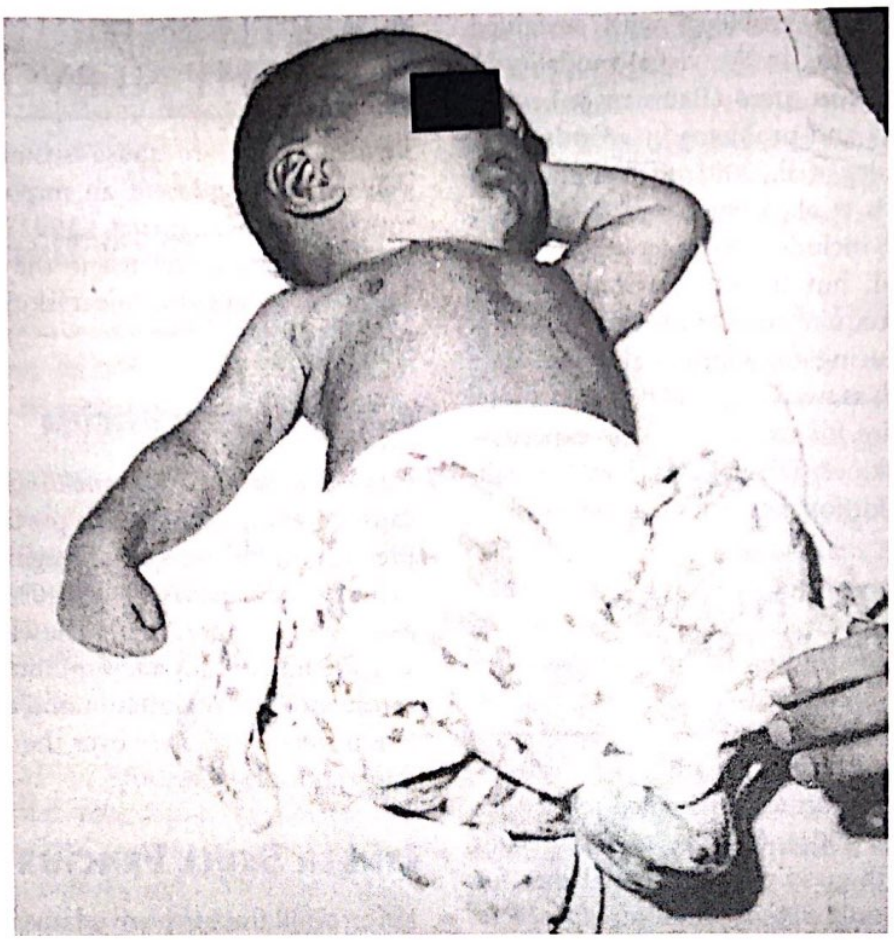
Caput succedaneum or molding is hemorrhagic edema caused by compression of a portion of the scalp from the pressure of the uterus or vaginal wall during a vertex delivery. Incidence is 20% to 40% of deliveries by vacuum extraction (Volpe, 2000). The clinical manifestation is a soft swelling usually a few millimeters thick. The edema is external to the periosteum and crosses suture lines. The lesion steadily resolves over the first days of life, and no intervention is necessary.

LINEAR SKULL FRACTURE

Linear skull fractures are relatively common in newborns and are caused by direct compression of the head during a prolonged labor and delivery and use of forceps during delivery. Uncomplicated linear skull fractures are diagnosed by radiographs and require no treatment. However, the fracture should alert the physician to the remote possibility of a more serious intracranial traumatic lesion (Volpe, 2001).

BIRTH BRACHIAL PLEXUS PALSY

Birth brachial plexus palsy (BBPP) is a paralysis or weakness involving the muscles of the upper extremity after mechanical trauma to the spinal roots of the fifth cervical (C5) through the first thoracic (T1) nerves during birth. Brachial plexus palsies are classified into three types: Erb's (73% to 86%), or upper arm, paralysis (involving C5 and C6); Klumpke's (less than 1%), or lower arm, paralysis (involving C8 and T1); and Erb-Klumpke (20%), or whole arm, paralysis (Dodds & Wolfe, 2000). Most cases of brachial plexus palsy follow a prolonged and difficult labor. The infant is often of high birth weight, sedated, and hypotonic, rendering the arm vulnerable to separation of bony segments, overstretching, and soft tissue injury (Mangurten, 1992; Scoles, 1992; Shepherd, 1991). Traction on the shoulder during delivery of the head in a breech presentation, and lateral traction of the head and neck while delivering the

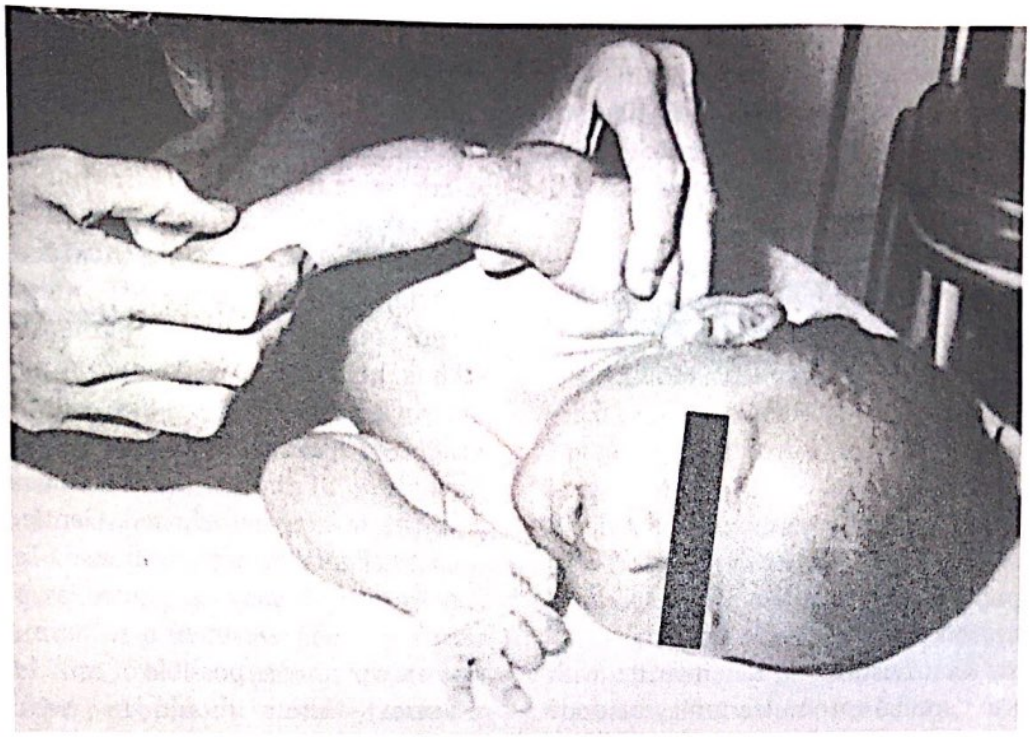


• **Figure 35-6** Four-week-old infant with partial paralysis of right arm as a result of brachial plexus injury. *(From Shepherd, RB. Brachial plexus injury. In Campbell, SK [Ed.]. Pediatric Neurologic Physical Therapy. New York: Churchill Livingstone, 1991, p. 107.)*

shoulders in a vertex presentation, can injure the C5 and C6 roots. More forceful traction may result in paralysis of the whole arm. Forceful elevation and abduction of the arm, stretching the lower plexus under and against the coracoid process of the scapula, may be the cause of the less common lower palsy. The birth trauma that injures the brachial plexus may also be associated with other lesions. Associated lesions may include injury to the facial nerve, fractures of the clavicle or humerus, subluxation of the shoulder, torticollis, or a hemiparalysis of the diaphragm by injuring the phrenic nerve at C4. The position of the arm and direction of forces that are applied are related to the injury (Jennett et al., 2002). It has been proposed that intrauterine fetal maladaptation is the cause of some neonatal brachial injuries with disuse osteoporosis as evidence (Jennett & Tarby, 2001).

In most infants with brachial plexus injury, the nerve sheath is torn and the nerve fibers are compressed by hemorrhage and edema. Clinical manifestations include a characteristic arm position of adduction, internal rotation with extension of the elbow, pronation of the forearm, and flexion of the wrist (Fig. 35-6). Passive abduction results in the arm falling limply. The Moro, biceps, and radial reflexes are absent; grasp is intact. Electromyography is used to detect a decrease in micropotential and signs of denervation. Magnetic resonance imaging can detect avulsions of the roots. Real-time ultrasound can be used to determine if the diaphragm is involved, which can occur with Erb's palsy (Volpe, 2001).

Treatment of brachial plexus injury includes rest for 7 to 10 days after injury to allow hemorrhage and edema to decrease. During this time, partial immobilization is accomplished by positioning the limb gently across the upper abdomen (Volpe, 2001). Physical therapy and occupational therapy begin after the initial period of immobilization and are important interventions throughout all phases of treatment of BBPP (Nelson, 2000). The goals of therapy intervention is to maintain range of motion (ROM) by gentle passive exercising, to stimulate muscle function as neural regeneration occurs, and to encourage active movement (Fig. 35-7) (see Shepherd, 1991, for a comprehensive physical therapy assessment and treatment plan). Splinting is controversial, and continuous splinting in the "Statue of



• **Figure 35-7** Therapist attempting to elicit activity in the deltoid muscle by encouraging hand-to-face movement. *(From Shepherd, RB. Brachial plexus injury. In Campbell, SK [Ed.]. Pediatric Neurologic Physical Therapy. New York: Churchill Livingstone, 1991, p. 112.)*

Liberty" splint is no longer recommended because it results in formation of abduction contractures and subsequent hypermobility of the shoulder. Intermittent splinting of the wrist and stabilization of the fingers are recommendations for lower arm palsy (Volpe, 2001; Mangurten, 1992). If improvement in active movement is not noted within the first few months, electromyography and nerve conduction studies are performed to determine the extent of damage, to follow recovery, and to determine if microsurgical repair is needed. Surgery is usually done at 3 to 6 months (Dodds & Wolfe, 2000).

Botulinum toxin injections into the antagonists of paralyzed muscles have been used in young children with brachial plexus injury to increase range of motion, decrease contractures, and improve body scheme (Desiato & Risina, 2001). It has been proposed that developmental dyspraxia may occur because of neonatal brachial plexus injury and resultant lack of, or inappropriate use of, the extremity (Rapalino & Levine, 2000; Brown et al., 2000).

SPINA BIFIDA

Spina bifida includes a continuum of congenital anomalies of the spine that range from failure of fusion of the posterior vertebral arch or arches to open spinal defects such as myelomeningocele (Edwards & Derechin, 1988) (see Chapter 25 for a full discussion of myelodysplasia). Approximately 80% of all lesions in infants with myelomeningocele occur in the lumbar area, which may reflect the fact that this is the last region of the neural tube to close (Volpe, 2001; Noetzel, 1989). Prenatal diagnosis of neural tube defects is based on levels of alpha-fetoprotein in maternal serum at 14 to 16 weeks, ultrasound, and acetylcholinesterase (AChE) assays. The sensitivity of AChE combined with real-time ultrasound in detecting neural tube deficits approaches 100% (Volpe, 2001). When an open defect is present, early closure of the back lesion is performed during the first 24 to 48 hours after birth to prevent infections such as meningitis or ventriculitis and to prevent trauma to the exposed tissue and stretching of the other nerve roots, which can lead to a loss of motor function. If prophylactic antibiotics are administered from 24 hours of life until surgery, infection rates are low. The most prudent course seems to be prompt closure after rational decision making by the parents and physicians (Volpe, 2001). The objective of sac closure is to replace the neural tissue into the vertebral canal, cover the spinal defect, and achieve a flat and watertight closure of the sac (Huttenlocher, 1987). An exciting new option is open fetal surgical repair of myelomeningocele at 20 to 26 weeks' gestational age with subsequent cesarean section delivery. The goals of open fetal surgical repair are to decrease trauma to the neural tissue, the need for shunting for hydrocephalus,

musculoskeletal deformity, and correction of Arnold-Chiari syndrome with the overall goal of improvement in neurologic function (Hedrick & Crombleholme, 2001).

Hydrocephalus is a common complication of myelodysplasia. The incidence of hydrocephalus varies according to the site of the lesion (Noetzel, 1989; Volpe, 2001). With lesions involving the thoracolumbar, lumbar, or lumbosacral areas, the incidence of hydrocephalus is approximately 90%. With occipital, cervical, thoracic, or sacral lesions, the incidence of hydrocephalus is 60%. Ventriculoperitoneal shunting is performed when there are clinical or diagnostic signs of hydrocephalus. Clinical signs include full anterior fontanel and split cranial sutures. Hydrocephalus is most commonly associated with overt clinical signs 2 to 3 weeks after birth. Serial computed tomography and ultrasonography are tools used to diagnose hydrocephalus.

Physical therapy examination of the neonate with myelodysplasia in the special care nursery may be done before or after closure of the back, or both before and after. This assessment is important to evaluate skeletal alignment, range of motion, motor and sensory function, reflex development, and behavioral organization. The neonate with myelomeningocele may have joint deformities as the result of imbalance of muscle action and positioning in utero. Positioning may also help improve or maintain skeletal alignment, such as prone positioning to maintain hip joint range of motion and encourage development of the extensor muscles. Side lying can be used to encourage a symmetric posture. The supine position should be avoided because of the effects of gravity. Splinting or serial casting may also be used to improve skeletal alignment (Schneider et al., 1995).

Examination of range of motion (ROM) in the newborn with spina bifida is indicated to identify impairments and, if necessary, to take advantage of the neonatal period of hyperplasticity of ligaments, resulting from transplacental transfer of relaxin and estrogen from the mother (Hensinger, 1977; Sweeney & Swanson, 2001). The therapist must be aware of normal neonatal physiologic flexion of the hips and knees and the possibility of hip dislocation. Because of the latter, it is recommended that hip adduction be tested only to the neutral position. Limitations in ROM are not an indication for aggressive stretching. ROM exercises should be performed slowly and gently to avoid fractures of paralyzed lower extremities. Common limitations in muscle extensibility for neonates with spina bifida include tightness of hip flexors, hip adductors, and dorsiflexors or evertors of the ankle (Mazur & Menelaus, 1991; Tappit-Emas, 1999). ROM exercises should be done with hands placed close to the joint being moved with a brief hold at the end of the range preventing unnecessary stress to soft tissue and joint structures (Tappit-Emas, 1999).

Muscle testing is performed by the physical therapist to determine the level of muscle innervation. Obviously, conventional methods of muscle testing are not appropriate for the neonate. Schneider and associates (1995) offer some strategies for eliciting muscle contraction that include carefully considering the state of the infant, tickling, holding the extremities in positions such as hip and knee flexion, and holding a limb in an antigravity position to stimulate the infant to move or hold a limb. Movements of the extremities may be observed, and contractions may be palpated. Gentle resistance may be given to elicit a stronger response (Tappit-Emas, 1999). Muscle strength may be graded as present, absent, or trace. Repeated assessment is recommended to obtain as accurate a picture as possible of muscle function.

Sensory testing should be performed when the neonate is in a quiet, awake state in order to determine the level of sensation. The skin is stroked with a pin or other sharp object to determine the neonate's reaction to pain, such as facial grimacing. The therapist begins at the anal area, the lowest sacral innervation level, and strokes across the buttocks, down the posterior thigh and leg, up the anterior surface of the leg and thigh, and across the abdominal musculature to determine reaction to pain (Schneider et al., 1995). The neonate's response should be recorded for each dermatome.

The physical therapist may also include reflex and behavioral testing such as the Brazelton Neonatal Behavioral Assessment Scale in the evaluation of the neonate with spina bifida (Brazelton & Nugent, 1995). The purpose of this part of the assessment is to evaluate reflex activity such as sucking and swallowing and to determine the current status of the infant's organization of physiologic response to stress, state control, motor control, and social interaction (Schneider et al., 1995). The physical therapist notes the neonate's strengths as well as the problems the neonate is having. Repeated behavioral testing may help monitor progress in organization and reflect recovery. When performing a reflex or behavioral assessment, it is important to ascertain whether the neonate's performance is affected by CNS-depressant drugs.

Intermittent taping, positioning, ROM exercising, and splinting are techniques used by the physical therapist to treat joint deformities. Intermittent taping is more adaptable to the nursery setting and is reported to be more effective than ROM exercises to improve mobility (Sweeney & Swanson, 2001). A thorough knowledge of arthrokinematic principles and techniques is necessary to

position the joint in a corrected position before taping. An external skin protection solution is applied under the tape, and an adhesive removal solution is used when the tape is removed. The therapist who performs taping must carefully observe skin condition and vascular tolerance when developing a taping schedule. Taping schedules begin with 1 hour and increase by 1 hour as tolerated.

PAIN

The neonate's ability to perceive pain has become a focus of both clinical and research attention (Pigeon et al., 1989). Previously, it was assumed that the neonate's nervous system functioned at a decorticate level, with insufficient myelinization of pain tracts and centers to perceive, feel, or remember pain (Franck, 1987; Shiao et al., 1987; Whaley & Wong, 1997). Increased knowledge of the capabilities of the newborn brain and advances in neonatal pharmacology have fostered a concern for the importance of protecting the neonate from stresses in the NICU environment. Evidence exists that pain pathways, cortical and subcortical centers of pain perception, and neurochemical systems associated with pain transmission are intact and functional in the prematurely born neonate. Slow conduction speed is the result of less myelinization, but it is balanced by shorter interneuronal distances traveled by painful nerve impulses and the fact that most nociceptive impulses are transmitted by non-myelinated C fibers (Anand & Hickey, 1987). However, the endorphin system, which mediates analgesia, may not be completely functional, leaving the preterm infant more sensitive to pain than term or older infants (Stevens & Franck, 1995). Infants may feel pain even more intensely than adults because of the immaturity of descending inhibition pathways in the spinal cord (Fitzgerald & Beggs, 2001; Anand, 1998; Franck, 1992). Early damage in infancy can lead to prolonged structural and functional alterations in pain pathways such as lower threshold and hypersensitivity that may persist into adult life (Fitzgerald & Beggs, 2001).

Although it is difficult to assess pain in the neonate, the physical therapist working in the special care nursery should be aware of the methods used to assess pain and be able to perform the nonpharmacologic procedures to alleviate pain. Both physiologic and behavioral responses of the neonate to nociceptive or painful stimuli have been identified. Physiologic manifestations of pain include increased heart rate, heart rate variability, blood pressure, and respirations, with evidence of decreased oxygenation. Skin color and character when pain is present include pallor or flushing, diaphoresis, and palmar sweating. Other indicators of pain are increased muscle tone, dilated pupils, and laboratory evidence of metabolic or endocrine changes (Chiswick, 2000). Neonatal behavioral responses to nociceptive input include sustained and intense crying; facial expression of grimaces, furrowed brow, quivering chin, or eyes tightly closed; motor behavior such as limb withdrawal, thrashing, rigidity, flaccidity, fist clenching, finger splaying and limb extension; and changes in state (Granau et al., 2000; Pigeon et al., 1989). An infant may, however, be experiencing pain when lying quietly with closed eyes (Shapiro, 1989).

Several neonatal pain measures have been developed, including the Neonatal Facial Coding System (Gruneau et al., 1990; Gruneau & Craig, 1987); the Neonatal Infant Pain Scale (Barrier et al., 1989); the Modified Behavioral Pain Scale (Taddio et al., 1995); and the Premature Infant Pain Profile (Stevens & Franck, 1995).

Nonpharmacologic methods to alleviate pain include decreasing stimulation, swaddling, nonnutritive sucking, tactile comfort measures, rocking, containment, and music (Abad et al., 1996; Burke et al., 1995; Franck, 1987; Whaley & Wong, 1997). Preterm neonates showed a significantly lower mean heart rate, shorter crying time, and shorter mean sleep disruption after heel stick with facilitated tucking (containing the infant with hands softly holding the infant's extremities in soft flexion) than without (Corff et al., 1995). Morphine, fentanyl, and topical mixture of a local anesthetic cream (EMLA) are the most commonly used analgesics in the NICU (Stevens & Franck, 1995).

FAMILY RESPONSE TO PREMATURE BIRTH

Although medical interventions are critical to the survival of the premature infant, there is an increased awareness of the importance of recognizing the needs of families and providing supports. Taking on the parenting role is a major life task that is greatly complicated by the crisis of a critically ill newborn. Families react to this crisis in different and individual ways. Common emotions include anxiety, guilt, fear, resentment, feelings of inadequacy, and anger (Berns & Brown, 1998). In addition, the high-tech NICU environment is unfamiliar, frightening, and stressful for parents and can interfere with parent-infant interaction (Miles, 1989; Sheilds-Poe & Pinelli, 1997; Ward, 1999; Willis, 1991). Als (1992) proposed that the normal emotional preparation for parenting is interrupted by preterm birth. The desynchronization of emotional unpreparedness compounded by fear for the life of the infant often leads parents

to experience feelings of helplessness, anger, grief, and sometimes prolonged depression. These experiences pose significant barriers to regaining confidence in oneself and daring to become invested in and committed to the infant. The costs to the other family members also must be considered. Als and colleagues (1992) suggest that goals for developmental care in the special care nursery include the following: (1) supporting parents as active partners; (2) helping parents learn how to observe their infant's stress and comfort signs; (3) assisting parents to develop competence in helping their infant to self-comfort, regulate, and organize behavior; and (4) reinforcing parents' feelings of their importance and effectiveness in caring for their infant. Parent education focusing on behavioral cues, handling, and positioning to improve parent handling and caregiving abilities is recommended as a goal of physical therapy (Unanue, 2002).

Although the NICU experience can pose a barrier to parent confidence, parents have rated themselves high in their abilities to perform caregiving activities and their understanding of information provided to them (Unanue, 2002; White et al., 2000). Parents should progress in the care of their infant at their own pace. Sources of formal support include staff, counseling, and parent group meetings. Coordination of care and parent education and support are essential at transfer and discharge time. Several studies report benefits of both hospital and home-based family-centered intervention after discharge (Cusson & Lee, 1993).

THEORETIC FRAMEWORK FOR NEONATAL PHYSICAL THERAPY

The theoretic basis for neonatal physical therapy may be conceptualized from general and specific frameworks. The model of the International Classification of Functioning, Disability, and Health (ICF) (World Health Organization, 2001) provides a standard language and framework for describing components of health and health-related states. High-risk neonates frequently demonstrate impairments in muscle tone, range of motion, sensory organization, and postural reactions. Limitations in activity may include problems in respiration, feeding, visual and auditory responsiveness, and motor abilities, such as head control and movement of hands to mouth. The interaction between impairments and activity limitations may contribute to restrictions in parent-infant interaction (participation). In the ICF model, the person and environment are contextual factors that influence body functions and structure, activity, and participation. Personal factors include an infant's health complications and temperament. Environmental factors include lighting and noise in special care nursery and family support. The examples provided potentially impact on all three components of health including heart rate, respiratory rate, oxygen saturation, feeding, and parent-infant interaction.

The purpose of the physical therapy examination and evaluation is to identify the presence of and risk for impairment, activity limitations and participation restrictions and contextual factors that may increase risk and determine appropriate interventions. Physical therapy intervention focuses on the needs of both the family and infant. Intervention includes handling, positioning, state transitions, and self-calming. In addition, as the infant approaches term age, developmental skills may be added to the intervention program. Another important focus of physical therapy intervention in the NICU is parent support and education. Parent education may include teaching parents about behavioral cues and how to identify their baby's cues, proper positioning and handling, and developmental activities to perform with their baby. Another role of the physical therapist is to determine the needs of the infants once discharged home. Infants may need to transition to community services such as an early intervention program or they may need to be followed through the NICU follow-up clinic on a consultation basis.

The American Physical Therapy Association, Section on Pediatrics has published guidelines for physical therapy practice in the NICU (Sweeney et al., 1999). These guidelines provide the physical therapist with a structure for clinical training, an algorithm for decision making, and clinical competencies based on roles, proficiencies, and knowledge areas. The NICU guidelines propose a theoretic framework based on the dynamic systems, the enablement and disablement model such as the ICF, preventive care, and family-centered care. The concepts within this framework optimize the functional posture and movement in infants to promote development. In the dynamic systems, all systems interact to produce a response (Thelan & Smith, 1994). When applying this concept to the NICU, the infant's biologic make-up, the sociocultural and physical environments, and the task interact to influence the function of the neonate.

Sweeney and Swanson (2001) have also described a theoretic framework specific to neonatal physical therapy that incorporates concepts of dynamic systems, neonatal behavioral organization, and crisis intervention. Neonatal behavioral organization concepts of this model

are those proposed in the synactive model of behavioral organization (Als, 1986). This model describes how newborn infants interact with the environment through five behavioral systems: autonomic, motor, state, attentional, and self-regulatory. The five systems are in continuous interaction with one another (synactive). The synactive model is useful when trying to determine the risk-benefit ratio for intervention, including the tolerance of a neonate to developmental examination and procedural interventions. For example, a neonate who reacts to examination procedures with autonomic and visceral stress signals such as gagging, tremors, or irregular respirations is demonstrating physiologic instability. These reactions indicate a higher risk than benefit from this developmental assessment. The neonate who reacts to the same assessment with smooth respirations, pink color, well-regulated tone, and smooth movements is tolerating the procedure with a beneficial interaction of subsystems. Determination of the risk-benefit ratio and tolerance of the neonate to developmental assessment and intervention is an important part of the theoretic framework proposed by Sweeney and Swanson (2001).

Physical therapists who work in special care nurseries should base their intervention strategies on current knowledge of neonatal development and intervention (Fetters, 1986). Models that address characteristics of the infant and the environment, such as the ICF and synactive model, provide a theoretic framework for the development of guidelines for intervention in the NICU. These models also allow for generation of hypotheses that may be examined through clinical research.

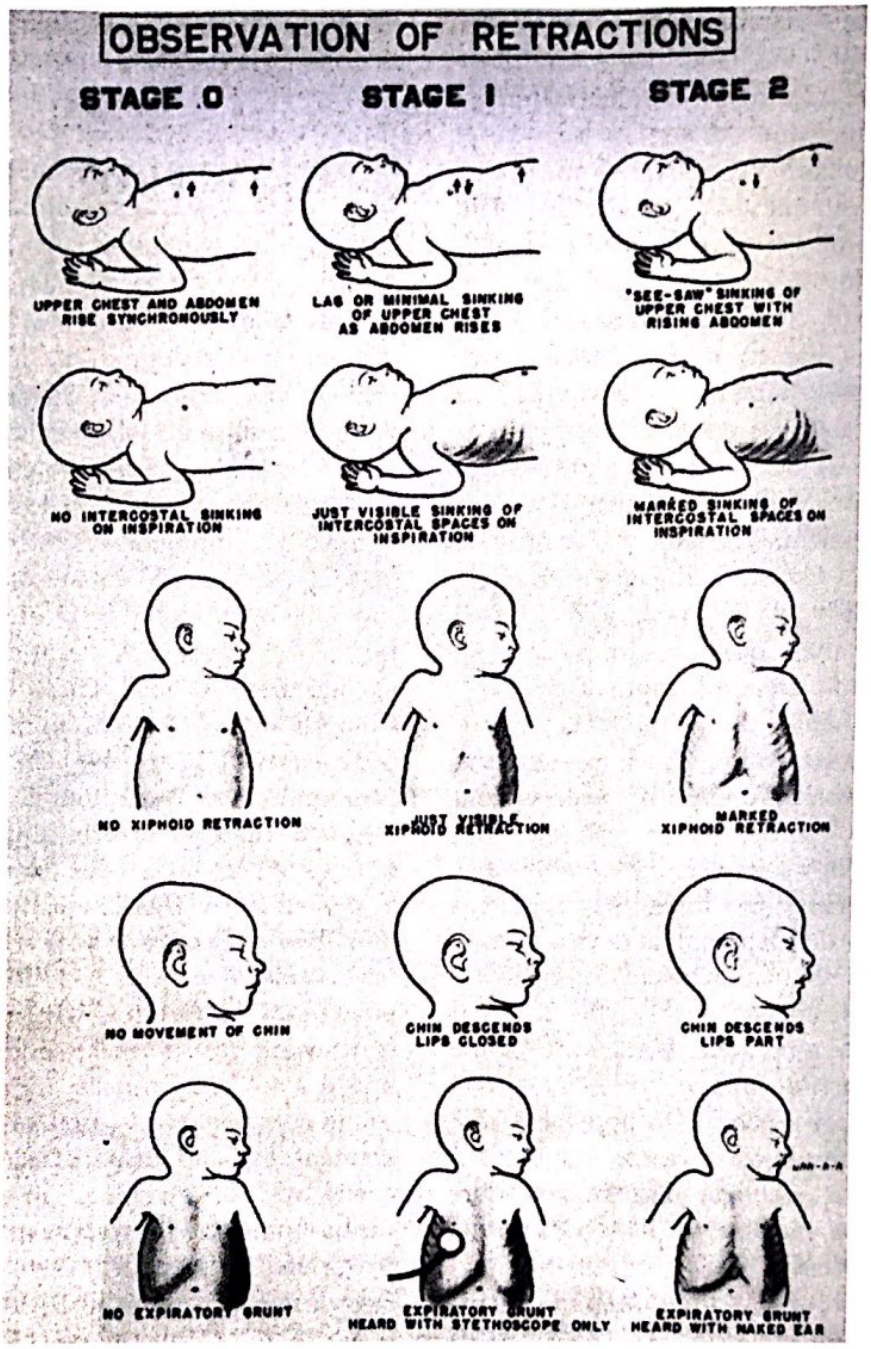
The physical therapist experienced in neonatal care has much to offer as a member of a developmental team in coordination of care, sharing information and providing instruction to families, prevention of impairments, and sensorimotor development. It has been recommended that physical therapists working in the NICU complete continuing education in neonatal medicine, fetal development, assessment and development in early infancy, parental education, early intervention, and interdisciplinary interaction in the specialized setting of the special care nursery (Campbell, 1985; Sweeney et al., 1999; Sweeney & Swanson, 2001). Knowledge of current theories of motor control and early motor learning is also beneficial. Other recommendations include a supervised preceptorship in an NICU, which may include training in the use of neonatal assessments such as the Neonatal Individualized Developmental Care and Assessment Program (NIDCAP) (Als, 1984) and the Neonatal Behavioral Assessment Scale (NBAS) (Brazelton, 1984). These assessments are discussed in the section on developmental physical therapy examination.

PULMONARY FUNCTION

Examination of pulmonary function in the infant requiring special care includes observation, inspection, and auscultation (Crane, 1995; Parker, 1985). Percussion is generally not appropriate for small infants (Crane, 1995). Observation and inspection include assessing signs of respiratory distress, chest configuration, skin color, breathing patterns, coughing, and sneezing. Signs of respiratory distress include retractions, nasal flaring, expiratory grunting, inspiratory stridor, use of accessory respiratory muscles (manifested by head bobbing), and bulging of intercostal muscles during expiration (Fig. 35-8) (Crane, 1995). Abnormal chest configurations include barrel-shaped chest and pectus excavatum. Cyanosis around the lips and mouth is a significant sign of hypoxemia (Crane, 1995).

Irregularity of respiration is normal in a neonate, and respiratory rates should be counted over a period of 60 seconds to account for this (Crane, 1995). The auscultation of an infant is an inexact procedure because of the thin chest wall, proximity of structures, and easy transmission of sounds (Fig. 35-9). This is also confounded by mechanical ventilation. During auscultation, the therapist is listening for normal, abnormal, and adventitious breath sounds. In infants, the specific location of the sound does not always correlate with the underlying lung segment. For this reason, auscultatory findings must be correlated with radiologic evidence. Palpation of the mediastinum and the trachea in the suprasternal notch is performed to ascertain if there is subcutaneous emphysema, edema, or rib fracture (Crane, 1995, 1987, 1986).

Positioning is indicated to enhance ventilation-perfusion ratios and to drain bronchopulmonary segments. Advantages of the prone position include improving oxygenation, lung compliance, state of alertness, and vital signs, whereas the primary advantage of the semierect position is to improve oxygenation (Crane, 1995; Martin et al., 1979; Wagoman et al., 1979). Positioning for postural drainage may need to be modified because of precautions and contraindications to certain positions in the neonate (Table 35-2). For example, the prone position is contraindicated by an untreated tension pneumothorax, and the head-down Trendelenburg position is contraindicated when there is an IVH of grades III and IV, acute congestive heart failure, or cor pulmonale (Crane, 1995).

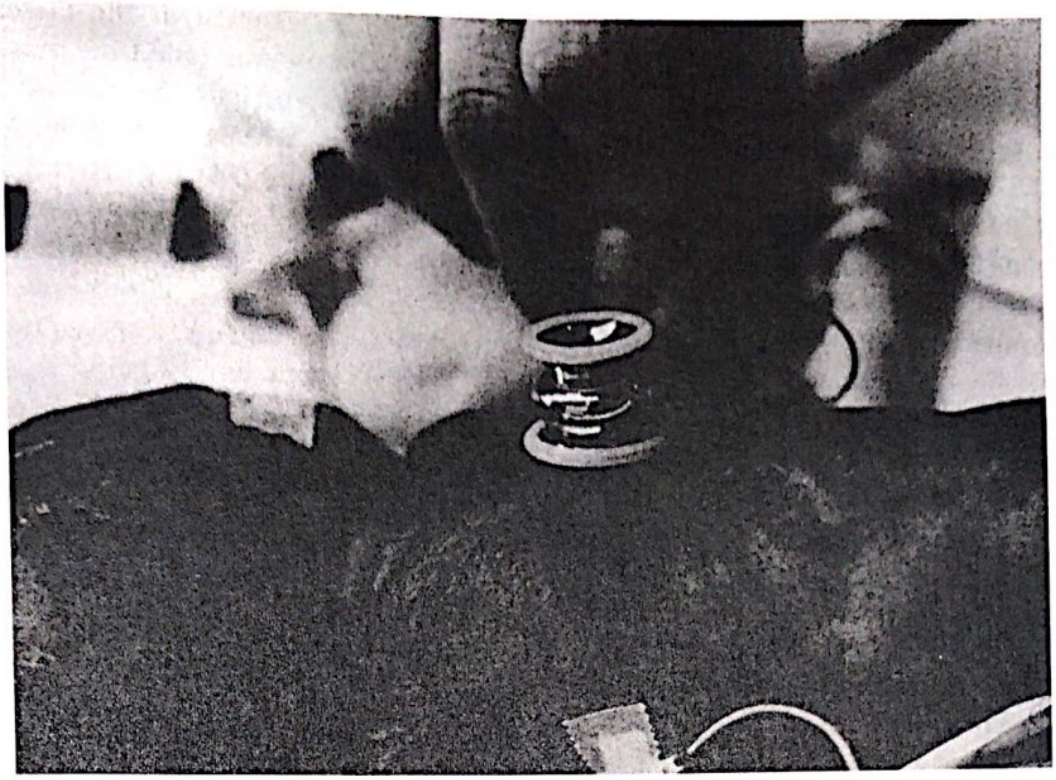


◆ **Figure 35-8** Observation and scoring of respiratory retractions. *(From Silverman, WA, & Andersen, DH. Controlled clinical trial of water mist on obstructive respiratory signs: Death rate among premature infants. Pediatrics, 17:1–10, 1956.)*

DEVELOPMENTAL EXAMINATION AND EVALUATION

The purposes of the developmental physical therapy examination and evaluation are to identify the following: (1) impairments in body function and structure that require intervention; (2) methods of positioning and handling; and (3) how to adapt the environment to optimize development. Based on examination findings, the physical therapist also evaluates the risk for activity limitations in development and parent-infant interaction. During examination procedures, the therapist must be aware of signs that the infant is undergoing stress, such as hypoxemia, excessive increase in heart rate, gagging, choking, and gasping. The therapist may need several brief sessions to fully assess the infant who is medically fragile (Sweeney, 1986). If the infant is too fragile for physical handling, observation of postures and spontaneous motor behavior may be useful.

Tests and measures appropriate for the premature infant include the Neurologic Assessment of the Preterm



• **Figure 35-9** Auscultation of infant's lungs with neonatal stethoscope. *(From Crane, L. Physical therapy for the neonate with respiratory disease. In Irwin, S, & Tecklin, JS [Eds.]. Cardiopulmonary Physical Therapy, 2nd ed. St. Louis: Mosby, 1990, p. 402.)*

TABLE 35-2 Precautions and Contraindications for Postural Drainage in a Neonate

POSITION	PRECAUTION	CONTRAINDICATION
Prone	Umbilical arterial catheter Continuous positive airway pressure in nose Excessive abdominal distention Abdominal incision Anterior chest tube	Untreated tension pneumothorax
Trendelenburg position (head down)	Distended abdomen SEH/IVH* (grades I and II) Chronic congestive heart failure or cor pulmonale Persistent fetal circulation Cardiac dysrhythmias Apnea and bradycardia Infant exhibiting signs of acute respiratory distress Hydrocephalus Less than 28 weeks of gestational age	Untreated tension pneumothorax Recent tracheoesophageal fistula repair Recent eye or intracranial surgery Intraventricular hemorrhage (grades III and IV) Acute congestive heart failure or cor pulmonale

From Crane, L. Physical therapy for the neonate with respiratory disease. In Irwin, S, & Tecklin, JS (Eds.), Cardiopulmonary Physical Therapy, 2nd ed. St. Louis: Mosby, 1990, p. 406.
*Subependymal hemorrhage/intraventricular hemorrhage.

and Full-Term Newborn Infant (Dubowitz & Dubowitz, 1981a, 1981b), the Neonatal Individualized Developmental Care and Assessment Program (NIDCAP) (Als, 1984), and the Test of Infant Motor Performance (TIMP) (Campbell et al., 2001). Tests and measures designed for at-risk full-term infants include the Morgan Neonatal Neurobehavioral Examination (Morgan et al., 1988).

Observation of movement is an important component of the physical therapist's examination and is included in many of the standardized assessments. The assessment of spontaneous movement in infants can identify infants who require early intervention services. Prechtl and colleagues observed videotaped assessments of movement in both preterm and full-term infants in order to identify infants in need of early intervention services (Prechtl et al., 1997). The presence of fidgety movements was a good indicator of normal neurologic outcome; abnormal and absent fidgety movements were indicative of poor outcome (Prechtl et al., 1997). Kakebeeke and colleagues (1997) developed a 10-point scale to rate the fluency and spatiotemporal variability of arm and leg movements and sequencing of general movement patterns. Significant differences were found for these three movement quality parameters among term, low-risk preterm, and high-risk preterm infants.

Neurologic Assessment of the Preterm and Full-Term Newborn Infant

The Neurologic Assessment of the Preterm and Full-Term Newborn Infant is a systematic, quickly administered, neurologic and neurobehavioral assessment developed by Dubowitz and Dubowitz (1981a, 1981b; Dubowitz, 1988). The test was developed to document changes in neonatal behavior in the preterm infant after birth, to compare preterm neonates with full-term neonates, to detect deviations in neurologic signs, and to document patterns of resolution in neurologic deficits (Sweeney, 1986). The test includes multiple neurobehavioral items of the Brazelton NBAS (Brazelton, 1984) using the six behavioral states. The neurobehavioral items include habituation to light and sound while sleeping, auditory and visual orientation responses, quality and duration of alertness, defensive reaction to cloth over face, peak of excitement, irritability, and consolability. The 15 neurologic items were taken from the Clinical Assessment of Gestational Age by Dubowitz and colleagues (1970), the Neurologic Examination of the Newborn (Prechtl & Beintema, 1964), and the neurologic examination of the full-term infant and the abnormal inventory protocol by Saint-Anne Dargassies (1977). The Neurologic Assessment of the Preterm and Full-Term Newborn Infant was tested on more than 500 infants during a 2-year period. The test takes 15 minutes or less to administer. Scoring is done by looking at patterns of response rather than by determining a summary or total score.

In a prospective study of the predictive validity of the Neurologic Assessment of the Preterm and Full-Term Newborn Infant, Dubowitz and colleagues (1984) found that 91% of premature infants who were classified as normal at 40 weeks of gestation were normal according to detailed neurologic assessment and the Griffiths Mental Development Scale at 1 year. Sixty-five percent of infants classified as abnormal at 40 weeks of gestation, including those with cerebral palsy, dystonia, global delays, clumsiness, and hearing or visual deficit, were considered abnormal at 1 year.

Although reliability has not been determined, this examination has great potential for physical therapists who work in the NICU because of its brief administration time and systematic method of recording. Caution must be used when administering this assessment because it has been documented that medically stable preterm infants demonstrated an increase in heart rate during and after this examination (Sweeney, 1986).

Neonatal Individualized Developmental Care and Assessment Program

The NIDCAP was developed to provide developmental observation and assessment training for neonatal health care professionals and includes Naturalistic Observation of Newborn Activity (Als, 1984). This involves systematic observation—without the observer manipulating or interacting—of the preterm or full-term infant in the nursery or at home during caregiving and handling. The NIDCAP is scored by observation and, therefore, is appropriate for infants who are unable to tolerate handling. A behavior observation checklist based on the concepts of the Assessment of Preterm Infant Behavior (APIB) (Als et al., 1982) includes environmental characteristics such as light, sound, and positioning apparatus. The self-regulatory ability of the infant is recorded, with some of the signs of stress being irregular respirations, color other than pink, tremors, startles and twitches, and visceral signs, including spitting up, gagging, hiccoughing, grunting, and gasping. Other signs of stress that can be recorded include flaccidity, frequent extensor movements of extremities, arching, tongue thrusts, finger splaying and fisting, fussing, yawning, and eye averting. Physiologic signs include heart rate below