PART 1

THE EXTREMELY PRETERM INFANT

Epidemiology, Perceptions, and Practices
INTRODUCTION

Three-year-old D is a vivacious small child who smiles and giggles freely. Her abdomen is criss-crossed with scars, the result of neonatal surgery for necrotizing enterocolitis for which she had surgical resection of some of her bowel. This was followed by the fashioning of an ileostomy that was closed at two years of age. There is also a scar over her left axilla, which followed a thoracotomy and the closing of a patent ductus arteriosus that had caused heart failure during the early neonatal period. She is the elder of twins, born at an uncertain gestation of 25 weeks weighing 810gs. Both babies were resuscitated at birth, but one twin died on day of life 4. Baby D received prolonged ventilation, required tracheostomy, and was discharged home on a ventilator after many months in the hospital. The daily nursing assistance the family received in their apartment was discontinued following the weaning of ventilation when the baby was aged 15 months. Her early years are remarkable for frequent visits to different specialists in the hospital who have monitored and managed her neurological development, pulmonary status, eyes, and gastrointestinal function. Her family, who have limited economic
resources, have undergone the most challenging of economic and emotional strains, and although they have faced the challenges most often with determined stoicism and love, there has often been anguish. However, D has conducted her only known life with the full gamut of emotional sparkle and oppositional irritation that would be expected from any able-bodied child. She has done this without the use of speech – a consequence of her tracheostomy and her profound deafness, the latter perhaps related to either her prematurity or aminoglycosides she received during the neonatal period. She is now a candidate for a cochlear implant, an option that would not have been available only a short time ago. She has started to use consistent sign to communicate, and her nonverbal developmental quotient is within the normal range.

Five-year-old B was born at 24 weeks’ gestation, weighing 580gs. At birth she did not breathe spontaneously, had a gelatinous feel to her skin, and could be held in the hand like a pound of butter. She was resuscitated and ventilated without much difficulty, and required about two weeks of intermittent positive pressure ventilation followed by a period of continuous positive airway pressure ventilation. She developed a grade II intraventricular hemorrhage, and following weaning from the ventilator there were many episodes of apnea and bradycardia, which responded to tactile stimulation. After 10 weeks in the hospital, she was bottle-feeding well and was discharged home one week later on an apnea monitor. During her early months, she was often an irritable baby who required frequent feeding, which was followed by episodes of regurgitation. The consequence of this gastroesophageal reflux was failure to thrive and choking episodes. The reflux failed to respond to medical treatment and after an admission to the hospital, because of severe aspiration pneumonia, she underwent a gastric fundoplication. Her irritability improved and she began to thrive. However, her development was relatively slow. She walked
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independently at 19 months and started to use two-word phrases at about three years of age. Her single-parent mother, who was aged 17 years at the birth of the baby, is now concerned and challenged because B demonstrates a reduced attention span, poor frustration tolerance, impulsivity, and emotional lability. These neurobehavioral difficulties have had an impact on her schooling, where she has difficulty staying in her seat, and with social interaction. Despite this, she is often a loving, affectionate child, with considerable charm. Psychometric evaluation was hampered by variable attention, but a minimum IQ level was measured at 86. There were some findings that suggested she may be at risk of demonstrating a specific learning disability, such as dyslexia, in elementary school. Despite her present difficulties, for which there are successful management strategies, and her extreme prematurity, she is expected to become an independent adult whose life will be governed by similar influences and fates that mould the outcome of any individual who was born normal at term.

J is a four-year-old boy who has recently started to walk using a walker. He is small, with relative undergrowth of the lower half of his body. He has a scaphocephalic head on which are perched thick glasses, and below these is an infectious open-mouthed grin, which is occasionally disfigured by a small amount of drooling. This, when he is reminded, is wiped away by an incoordinated splayed hand. He loves to demonstrate his walking ability and can hurtle down a corridor, albeit in an ungainly fashion, with hips and knees bent and knees knocking and on his toes. This is accompanied by much mirth shared by J and his onlookers. He is adored by his parents and two older sisters, and he adores them. J was born at 24 weeks, weighing 610gs. He required several weeks of artificial ventilation and developed a grade III intraventricular hemorrhage and pronounced periventricular leukomalacia. He required gastrostomy feeding for the first two years of his life, and he has had surgery
for retinopathy of prematurity and for a strabismus. His cerebral palsy and poor development was apparent during the first year of life, and repeated cognitive assessments place him in the mild mental retardation range with non–gross motor developmental quotients ranging from 60 to 70. He is expected to achieve adulthood and live a life that, although requiring some assistance and protection by others, will be one in which he is competent in the activities of daily living and able to benefit from some basic education and training.

T is aged five years. He was born at 25 weeks' gestation, weighing 700gs. Resuscitation was achieved easily after birth, and he was ventilated with relative ease for about three weeks. On day of life 5, he had developed a grade IV intraventricular hemorrhage, which was accompanied by severe periventricular leukomalacia. By one month of life, he was breathing independently but was unable to feed and would later require a gastrostomy. It was soon clear that he would develop substantial neurological handicap. Severe spastic quadriapresis, anarthria, pseudobulbar palsy, microcephaly, and what appears to be severe mental retardation now confine him to a wheelchair. He is unable to feed himself and continues to be fed by gastrostomy. He is incontinent and cannot indicate his needs. However, he appears to respond to familiar voices and smile socially and laughs with his siblings. Successful voluntary movements are not possible, and any stimulus or attempt at movement invokes mass, uncoordinated, stereotypic postures. There are contractures in his arms and legs that hamper dressing, toileting, and hygiene.

These cameos are very familiar to anyone involved in neonatal care and follow-up. They represent some of the complications of prematurity, which vary in their severity and cause considerable individual, social, and economic burden. Although it is the
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severely disabled child that may be most readily remembered, this outcome is not the rule. However, all adverse outcomes become more likely as birth weight and gestation decrease. Survival rates for low birth weight and preterm infants are giving rise, it appears, to an increasing prevalence of childhood neurodevelopmental disability, including severe forms of cerebral palsy. This has raised bioethical and legal questions concerning this population of children. These include topical and debatable concepts such as the limits of viability, end of life decisions for those without capacity, futility, parental and physician autonomy, distributive justice, the role of statutory and case law, and so on.

For the purposes of this book, I define the extremely preterm infant (EPTI) as one who is born at less than 28 weeks’ gestation. I also include the extremely low birth weight (ELBW) infant born weighing less than 1,000g. The two are not synonymous as the latter may include infants who are small for gestational age and more mature than the former. However, the literature includes both groups, and for the purposes of argument I do the same.

Extreme prematurity is uncommon, occurring in about 1% of live births(1). However, the moral dilemmas that arise from intensive care for EPTIs is a continuing cause for concern. Although, for some attitudes are fixed, for many the situation is fluid. But the question remains the same. How far should those go, who care for children, to preserve life at the inevitable expense to some babies, families, and society of disability, emotional trauma, and financial cost? Furthermore, attempts to answer this question are clouded by uncertainty arising from the limitations of early prognosis, variable and changing results of management, and differing subjective judgments from health professionals, parents, guardians, and the creators and arbiters of the law. Attempts to resolve the
conflict are sought from religion, bioethics and moral philosophy, sociocultural acceptance of certain behaviors, and the law, both civil and criminal. But before these can be considered, it is necessary to briefly provide some history and then document the epidemiology of EPTIs, the perceptions of those involved in their care, and the resources expended.
HISTORICAL ASPECTS

Depending on cultural, religious, and socioeconomic circumstance, infanticide occurred throughout history. (2,3) During the classical period, infants deemed abnormal were left to die in the open, (4) and infanticide was not unusual up until the 20th century. (5,6) But as medical expertise and technology have become increasingly sophisticated, active measures are now taken to keep alive such infants, and the degree of this endeavor has mirrored changes in societal attitude. This is particularly evident for the EPTI. However, the requirement that physicians should not provide treatment that they believe will be of no benefit can also be dated back to the classical era, and there may well be a positive obligation not to do so. Hippocrates wrote that: “[W]henever a man suffers from an ill which is too strong for the means at the disposal of medicine he surely must not expect that it be overcome by medicine,” and, he continued, for the physician to provide treatment in such a situation was “allied to madness.” (7)

And Plato, in The Republic, advised that the physician should:
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For those whose bodies were always in a state of inner sickness he did not attempt to prescribe a regime . . . to make their life a prolonged misery . . . medicine was not intended for them and they should not be treated even if they were richer than Midas.(8)

Out of this history has arisen a requirement to care for the EPTI, but not to oblige a physician to provide treatment that is perceived as not beneficial. However, because of differing beliefs, perceptions, and interpretations, there may be a conflict between the requirement and the obligation.
From 1980 to 2000, the infant mortality rate in the United States has been reduced from 12.6 to 6.9 per 1,000 live births. (9, 10) This has occurred with an approximately 17% increase in preterm birth rates, (9, 11) and reductions in mortality have been highest for those with the lowest birth weights. (9) This has been mainly attributable to gains in technology as well as improvements in medical practice. (12–15) ELBW infants account for nearly half of total perinatal mortality, despite being only a very small percentage of total live births. (16) Much of the improvement in mortality has occurred in the very and extremely preterm groups. (17, 18) There can be considerable variation in the results of studies reporting mortality for the EPTI. To some extent this is governed by the conduct of the studies, (19, 20) for example, whether the figures reported include total births, live births, or neonatal intensive care unit (NICU) admissions; whether the numbers were small or based on geographic populations; and whether there were consistent approaches to management. Clearly this variability may introduce uncertainty and incomprehensibility into the counseling of parents. Furthermore, one can speculate