
The Quality of Life of Young Children and Infants with Chronic Medical Problems: Review of the Literature

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The question “what makes a good quality of life?” is a philosophical one which could be thought immune to scientific investigations. However, over the last few decades there has been great progress in developing tools to quantify quality of life (QoL) to make comparisons between different health states, evaluate the effectiveness of medical interventions, and describe the life trajectories of individuals or groups. Using a series of vignettes, we explore and review the biomedical literature to demonstrate how QoL is affected by chronic health conditions in childhood, and how it evolves as individuals pass into adulthood.

Individuals experiencing serious chronic illnesses generally have reduced health-related QoL: their health status has significant repercussions of their everyday life, but scores are usually

much better than healthy individuals expect, and better than physicians predict. Global QoL is more than a health status concept. QoL is a complex relationship between objectivity and subjectivity; it requires substantial and valid facts, and it defines itself by an interpretation of health within different schemes of values: societal, medical, and those of the subject themselves. QoL is dynamic; purely physical influences diminish as individuals age, and psychosocial factors become much more important. Resilience frequently allows adaptation to adverse health states, leading to acceptable QoL for most children with disabilities.

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Quality of life (QoL) is a subtle construct which necessarily implies multiple concepts of very differing nature; it requires objective facts, determined by social relationships, physical health, and physical and mental abilities; and subjective experiences, relating the individual’s capacities to the environment in which they live, implicating factors, such as the degree of independence, a sense of well-being, and personal values. Health-related quality of life (HrQoL) is a somewhat more restricted concept, focusing on the repercussions of one’s health-status on the meaning of one’s life, in terms of psycho-

logical, social, and physical well-being. QoL therefore has 3 characteristics: it resides within a complex relationship between objectivity and subjectivity; it requires substantial and valid facts; and it defines itself by an interpretation of health within different schemes of values: societal, medical, and those of the subject themselves. It is commonly stated that “quality of life” and particularly HrQoL are the most important outcomes of health care interventions, and to analyze the benefit or risk of a particular intervention, QoL/HrQoL outcomes should be measured and compared.

The family of a severely limited young infant may evaluate their QoL very differently to the population who have never been in that situation

Although perhaps conceptually QoL is primarily qualitative, many tools have been developed in an attempt to quantify it, to evaluate and compare QoL in different circumstances. Constructing such tools is in itself complicated; what weight is given to objective facts as opposed to subjective experience? A life with severe functional limitations may be considered of high quality by the individual themselves, but of very

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limited quality to external observers.¹ The family of a severely limited young infant may evaluate their QoL very differently to the population who have never been in that situation, as clearly demonstrated in moving and sometimes painful detail by the narratives in this issue. Why do individuals in situations of severe functional limitation often evaluate their own QoL so much more positively than independent observers? Perhaps it is because in some circumstances the alternative for the individual is to be dead (such as seen in studies of home ventilation among children with life-shortening illnesses²), or because the individual has never experienced another kind of life (such as seen in home ventilation studies which include children ventilated since infancy³). More commonly it seems that this discrepancy, or paradox, as it has been called,¹ arises because individuals experiencing chronic health problems find value in things that observers without those limitations value less⁴; they also may find fulfillment in overcoming their limitations, and in helping others to adjust to their problems.¹

Tools for Measuring QoL in Children

A systematic review of HrQoL studies published in 2001⁵ found 137 articles describing QoL measurement in children with chronic diseases. They identified 22 different scoring systems for generic usage, and 21 designed for specific health conditions. They considered that each scale had limitations, and that certain underlying assumptions regarding QoL research in children still needed to be addressed. Many of these concerns remain, although some scales which have been updated more recently have addressed many of the problems. One well-supported scale, for example, the Pediatric QoL inventory, has itself been used in over 400 published studies since 2001.⁶

If evaluation of QoL and HrQoL in the face of known and measurable deficits in health status is difficult, *prediction* of QoL, performed during the neonatal period, is much more so. In addition to the uncertainties associated with how a life might be subjectively evaluated are added the uncertainties regarding whether an individual may or may not develop objective functional limitations.

In certain circumstances functional limitations are very likely but of uncertain severity, for example, in infants with structural anomalies, such as meningomyelocele. In other circumstances, particularly for the very preterm

infant, it is often very uncertain whether an individual will have any limitations at all, minor limitations, or serious handicap. In still other conditions (such as hypoplastic left heart syndrome) research has until recently focused exclusively on survival, and both functional and QoL outcome studies are scarce.

In addition to these concerns are other specific issues related to the young infant: how are factors, such as behavioral problems, added to the mix? The individuals themselves may not perceive that they affect QoL at all,⁷ but they may be considered to have major effects on HrQoL by parents.⁸ Can such difficulties be addressed in QoL scales? If so, with what weight?

Tools that examine HRQoL can be divided into direct and indirect measures. When direct tools are used, the respondent evaluates a health state, often on scales ranging from 0.00 (death) to 1.00 (perfect health). These tools can be used to evaluate either a hypothetical health state or the respondent's own health state. Such direct methods include the visual analog scale and standard gamble techniques. In indirect measures, the respondent provides information regarding their health status by completing a questionnaire. The responses are then converted to a score from a formula typically based on preferences from a sample derived from the general population sample. This inclusion of the preferences of the general population in the calculation of scores from indirect measurements is thought to be one reason direct and indirect measurements when completed by the same individuals differ widely.⁹ Indeed, it has been questioned whose values are the most important in determining health care decisions.¹⁰ Patients who have actual experience of a health state can perhaps better assess its effect on their HrQoL and express a true preference. The general population, by contrast, may be considered more objective. However, often, fear of a given health care outcome may be based on prejudices, with very little actual experience of the particular outcome. Such prejudices and fears may be a bias that makes healthy individuals rate illnesses, particularly chronic illnesses, much worse than those who live them.¹ This is not because chronically ill individuals rate all chronic illnesses less negatively than the population as a whole; it is a phenomenon specific to their actual condition.¹¹

Even though QoL studies have been criticized, it seems to us that knowledge of how individuals experience their own lives is essential as a means to evaluate the usefulness and success of interventions. The fact that even impaired survivors can view their lives in a positive light has been dismissed as "making lemonade" by some

commentators.¹² We feel, in contrast, that it is a cause for celebration that individuals with impairments (lemons) can nevertheless find value and meaning in their lives (and make lemonade)? Such resilience should not be dismissed, but should not at the same time impede our efforts to investigate methods to reduce such impairments. Even though subjective HrQoL may be acceptable to even those survivors with substantial medical problems and permanent functional limitations, efforts to reduce such problems will improve objective QoL, reduce long-term costs to medical systems, and make life easier for survivors and their families. For example, survival with serious visual limitations may be considered acceptable to those who are in that situation and coping with their blindness; such individuals will likely have good scores on QoL scales, but interventions to reduce visual impairment would still be welcomed by anybody, the visually impaired as well as those with normal vision.

We can illustrate the gulf which sometimes may exist between subjective and objective evaluations of QoL with studies regarding the QoL of infants with meningomyelocele, who have multiple functional problems in the long term, but have a QoL which they themselves find to be good or very good, including those who are wheelchair bound and incontinent.¹³ Questionnaire studies of healthy adults in contrast assigns very low health utility scores to being wheelchair bound, or being irreversibly incontinent.¹⁴ Interestingly, this appears to be a universal psychologic response to adversity; when a condition is irreversible, the resilience of the individual and adaptation to the new reality improve well-being, leading to HrQoL, which is only mildly affected. In contrast, the same chronic adverse health states, when they are potentially reversible, significantly reduce HrQoL. This paradox has been cleverly investigated by Smith and colleagues,¹⁵ evaluating a single specific health care problem, a new colostomy, in 2 samples of patients: those in whom it was known from the start that the colostomy would be permanent, and another where there existed a possibility of reversal in the future. Six months following the procedure, those with a known permanent colostomy had a substantially better HrQoL than those in whom the long term was still uncertain and the possibility of reversal persisted.¹⁵ This has been described as being “happily hopeless,” a phrase which succinctly embodies the paradox, which upon reflection is not so paradoxical. When there is no hope of reversal, the individuals find ways to cope, adapt to

the limitations and difficulties that their new situation demands, and find ways to incorporate their changed health status in their daily lives, and as a result their attitudes change.

Quality of Life in Selected Conditions in Childhood

We concentrate on 3 conditions for which intensive interventions in the neonatal period are frequently required to maintain life and improve function: extreme prematurity, myelomeningocele, and hypoplastic left heart syndrome.

In each case a varying amount of literature exists that can be used to illuminate the QoL of survivors. One could question, what is the purpose of such studies? According to Guyatt et al,¹⁶ QoL studies can be used to discriminate between individuals or groups, predict or classify individuals into categories, or evaluate longitudinal changes in an individual. We would add that QoL can be used as a primary outcome in prospective randomized trials.

To demonstrate the complexity of assessing QoL for an individual, we present examples of each of the conditions listed above to illustrate the notion of QoL, what it can mean and how it evolves, through published literature.

Extreme Prematurity

Before

Lucy is only in the 23rd week of pregnancy when she arrives at the hospital with severe pre-eclampsia. She may need to be delivered rapidly if her condition deteriorates. At this point, still under the terrible distress, she and her husband Marc meet with the obstetrician and with the neonatologist. They hear terrible things about their baby, Zack. Nobody can be sure that he will survive, but they are told that even if he does, he would have a substantial chance of having neurological damage, being handicapped, and having a many serious problems during his life. After 2 nights of insomnia, Lucy's medical situation worsens and she is told that she needs to be delivered as her own health is in danger. Confused and unsure of what they are doing, Lucy and Marc decide to opt for a Caesarean section as they understand that it would at least give their little boy a chance.

Birth

Zack takes his first breath on January 18 at 2 AM. He is just 24 weeks gestation and weighs 640 g. He does not move much and is immediately removed from his mother; a few seconds later he feels a cold metallic blade going into his mouth and a tube being inserted in his throat. He is then taken to neonatology where there are a few very busy hours, with multiple interventions, including insertion of central catheters. If he makes it, he might go home in 4-5 months.

Neonatology

Zack starts his long journey in neonatology. Initially all goes better than average, but a few days after birth, he has a serious deterioration, with sepsis. He deteriorates quickly, requiring several episodes of resuscitation for respiratory arrest; he develops shock and requires infusions of dopamine to maintain tissue perfusion. In addition his ductus arteriosus has reopened and worsens his clinical state. In the course of this acute deterioration, it becomes clear that he has necrotizing enterocolitis with a bowel perforation.

He is in a critical condition. He no longer looks like the same baby; he hardly looks like a baby at all: gray, marbled skin, swollen with edema. Marc and Lucy are devastated; everything seemed to be going so well and this deterioration is the worst thing that could have happened, even though it was half-expected.

When they visit the unit, they hear the nurses asking whether it is reasonable to continue intensive care. They suddenly feel on the edge of the precipice. The doctors try to be more reassuring, pointing out a few little signs which are encouraging, opinions which seem so different from the nurses. Are things being hidden from them?

Finally Zack recovers. After further complications of lesser severity, he develops bronchopulmonary dysplasia requiring oxygen therapy at home, where he goes 4 weeks after he was due to have been born.

At Home

Early Infancy. Lucy and Marc, who are excited to have him home, are already exhausted. They are very apprehensive about their first months with Zack. Lucy has already used up most of her maternity leave and will have to resign from her job to give Zack the on-going care he needs. She won't even consider a nanny or day care; he is far too fragile. The first months are very difficult; Zack does not eat well and

gains less weight than the pediatrician wants. His first developmental examinations show many areas of concern: his cognitive development is slow, and he is falling off the growth percentiles. The pediatrician worriedly suggests physiotherapy to stimulate his motor development. In addition, the difficulty in getting a good response from his hearing screen leads to repeated testing, which confirms that he is deaf in 1 ear. A hearing aid is prescribed to help his speech development, but he hates it. He does not walk until after his second birthday and is quite unstable.

At times the problems seem endless. Lucy is often at the hospital with Zack, for follow-up visits or, twice during his first year, for hospitalization for a chest infection. Marc often comes home late, and the couple has much less money since Lucy gave up her job. They have given up the idea of another child. These problems lead to difficulties between the couple, and they almost split up more than once. When they watch Zack's progress, however, he brings them much joy. He is a little boy who is timid, curious, and charming. He loves his father to take him on his lap to tell him animal stories. He clings to his mother who he watches always from one corner of his eye. Even with all these effects on the function of the family, they realize each day that their little guy is growing, bravely facing his problems, and bringing them happiness.

At School

At 5 years of age, Zack is enrolled in school. He is frightened and does not want to let his mother leave. He has had few contacts with infants his own age and never been to day care. His integration is difficult; he is solitary and sad. The teacher is concerned. Zack seems to have good understanding but does not participate and rests apart from the other children. He often seems to be dreamy, looks at his books, and has difficulty concentrating.

The teachers wonder about his intelligence; he performs less well than the others, but it is not because he has major behavior problems. Marc and Lucy want to help him. They would like to get him extra tutoring but they have limited means. He has to repeat his second year and is with a group of children who are younger. It seems to help him; he is better adapted now and enjoys school more and progresses.

Adolescence

At secondary school Zack has fewer problems. He makes new friends who are more interested in him

than his hearing aid and don't make fun of his limp. He is happier and starts to make plans for his future. Marc and Lucy remain very attentive to his needs, even if Lucy has restarted to work, and Zack has more interests. He has problems participating in many sports; his left leg is stiff, because of cerebral palsy, and he cannot run with his friends. He has become more popular, with his gentle and thoughtful demeanor. He still has difficulty with academic pursuits, and he has difficulty gaining his school leaving certificate. The day of his graduation Lucy and Marc are so proud they have tears in their eyes.

Objective Developmental Assessment. Zack suffers from 2 major disabilities secondary to his extreme premature birth and hospitalization. He is deaf in his right ear, needing a hearing aid, and he suffers from mild spastic monoparesis (cerebral palsy). During his school years, he also "suffers" from a form of attention deficit disorder as well as an intellectual capacity in the lower part of the normal range (his IQ is 85).

Prediction of QoL by Obstetricians, Pediatricians, and Caregivers. Zack's life starts with a high risk of not beginning at all. Parents are exposed to an enormous amount of information, much of it negative, leaving little space for hope and for their projected life as a family with their son.¹⁷ Information is mostly "medical" and based on prognosis for functional outcomes. However, it is well known that physicians underestimate survival rates from 23 through 34 weeks' gestation and freedom from serious handicap from 23 through 36 weeks' gestation.¹⁸ They advocate early treatment to try to suppress preterm labor, but less than 50% would perform Caesarean delivery for fetal distress before 26 weeks' gestation.¹⁹ Among physicians, even pediatricians expect less favorable outcomes for premature babies than published national rates of survival and freedom from handicap.²⁰

With such a negative framework, it is difficult to imagine that one could anticipate good or even fair QoL. Even on a subjective level, evaluation of QoL should include both the positive and the negative aspects of life. However, it seems that "informed consent" as valued by the medical profession emphasizes the negative aspects of health for premature babies.¹⁷ Indeed, statements of professional societies give a great deal of guidance on how to inform parents of the frequencies of complications, and of long-term health and developmental problems. We are urged to list all the potential significant complications in the short, medium, and long term, ensure by asking

questions that the parents have well understood the message, give percentages or other ways of indicating the frequency of various complications, and be sure to clearly describe what cerebral palsy and a reduced developmental quotient really mean.²¹ We question whether most neonatologists themselves understand the implications of these diagnoses well enough to explain them to parents and contrast these directions with the long-term experience of former preterm infants and their parents who almost universally rate their QoL as being acceptable, good, or excellent.²²

These considerations are of critical importance, as many decisions to withhold or withdraw treatments are taken during this period, when projected QoL is uncertain, and decisions are taken by proxies (parents and caregivers), who often have little to go on. Indeed, systematic reviews are unable to clearly show that HrQoL is correlated with gestational age at birth.^{23,24} As a result, one might ask, if HrQoL is unrelated to gestational age at birth, surely the only ethically appropriate factor that should be included in decisions to intervene or not is the probability of survival? If that is agreed, then should we not apply similar criteria for acute care treatment of all critically ill patients, regardless of their age? We have shown that different standards are applied to such decisions in the neonatal period²⁵: in a questionnaire study, respondents said they would be more likely to resuscitate an adult with far worse predicted outcomes than a preterm infant, and much more likely to accept a family's request for palliative care for the preterm infant.

After birth, different caregivers value different things about the child. Physicians and nurses are relatively similar in their projection of QoL.²⁶ However, during the hospital stay, nurses seem to attach more on observation and "gut feeling" about their projection of the newborn taken care of.²⁷ Nevertheless, they strongly differ from parents, who significantly project a better QoL.

Infancy. As reviewed by Saigal and Tyson, infancy is marked by a lower QoL for children and parents,²⁸ affecting all aspects of life, such as health status and behavioral and anxiety problems.^{29,30} Differences are most notable in motor functioning.³¹ Interestingly, as an evaluation by proxy, mothers' well-being seems to correlate with their perception of their child's QoL.³² This is probably due to the impact of health problems in early infancy for ex-premature children and multiple hospitalizations. In addition, premature birth may cause considerable stress for parents and result in post-traumatic stress disorder symptoms, such as in-

vasive memories, attempts to avoid or ignore certain specific experiences, and emotional vigilance. This may have implications with regard to the transition to parenthood, namely on parents' representations and care-giving competencies.³³

School Age. Interestingly, there is little literature regarding school age QoL of ex-preterm children. Saigal et al³⁴ demonstrated that HrQoL was significantly lower for extremely low birth weight (ELBW) children compared with the general population. However, this evaluation was not made from children or their parents. One study showed that parents tend to be much more protective of children at the age of 8.³⁵ This indirectly indicates that some of these children are still an important source of worries for their parents.

Adolescence. Arriving at adolescence, Zack performs a little worse than his peers. He still needs his hearing aid and has some difficulties running, his leg being a little stiff. However, he develops a social network and is able to follow the regular scholar curriculum.

ELBW teenagers report significantly lower utility scores than their peers, but not necessarily a lower HrQoL.³⁶⁻³⁸ Similarly, very low birth weight (VLBW) teenagers did not rate themselves as significantly different from their peers on a generic health measure. Adolescents of extremely low gestational age (ELGA) infants were significantly more limited because of physical health issues.

In contrast, parents of these teenagers reported significantly lower scores in their child's behavior compared with parents of children in the control groups. Parents of VLBW teenagers with low IQ indicated that their children performed significantly lower in terms of global health and behavior, general health perception, self-esteem, and family activities. They reported significantly lower QoL.²⁴ Parents also reported a significant impact on their own psychosocial health, mostly notably in emotional impact and time, regardless of their child's IQ.

From a psychosocial standpoint, ELBW teens reported lower scholastic, athletic, job competence, and romantic confidence and viewed themselves as more likely to need help from others in finding a job. In the behavioral domain, parents reported their ELBW teens to display more internalizing, more externalizing, and more total problems than the control teens, with ELBW boys showing more problems.³⁹ However, their self-esteem is similar to those born at term. This is particularly interesting as chronically

low self-esteem may contribute to anxiety, depression, lack of motivation, and poorer achievement in both occupational goals and interpersonal relationships at adulthood.⁴⁰

Young Adulthood. Transition from adolescence to young adulthood is a difficult time for Zack and his parents. It means a beginning of independence for a child who has always needed more support than the others. Some overprotection³⁵ during childhood may even complicate such a transition. Transition to adulthood for individuals with disabilities may be considerably delayed because of physical limitations, cognitive deficits, educational challenges, fewer opportunities for social integration and job market prospects, and a lack of acceptance by peers and employers.⁴¹ Although many individuals with disabilities may be able to achieve some of the expected roles, a minority of very severely disabled individuals may continue to be dependent on their parents or siblings lifelong.⁴²

Compared with their peers, there were no differences in QoL or subjective QoL as reported by young adults who had been born at VLBW.^{43,44} Saigal et al also showed comparable scores on HrQoL between ELBW and their peers.²²

Socially, young adults born ELBW are only slightly disadvantaged regarding peer and family relationships, friends, participation in sports, clubs, driving a car, social drinking, romantic relationships, and sexual experiences.⁴⁵⁻⁴⁷ No differences have been reported between the premature group and normal birth weight (NBW) group in their satisfaction with life, self-esteem, and behavioral functioning, except for a slight tendency to increased risk of depression among girls.⁴⁸

IQ. It is well recognized that IQ is highly correlated with parental socioeconomic status and educational background as well as environmental factors, both positive and negative.⁴⁹ It is simplistic to believe in the long-standing cause-effect inference model in which, for example, being born at ELBW is assumed to have a linear impact on cognitive and neuropsychological outcomes years later.⁵⁰ Socioeconomic gradients have a powerful effect on cognitive and behavioral development from infancy to childhood and beyond.⁴²

Resilience. Rutter⁵¹ brought to our attention the need to focus on protective processes that could exert their influence to bring about changes in life trajectories from risk to adaptation. In the longitudinal studies from the island of Kaua'i of multi-racial children with

cumulative effects of perinatal stress, poverty, parental psychopathology, and chronic discord, Werner⁵² identified several clusters of protective factors and processes, linked over time, which enable vulnerable individuals to escape from adversity and contribute to positive outcomes in adult life. These protective factors could be inherent in the children themselves, or within the family or community. Werner⁵³ and Smith's¹⁵ observations support the concept of resiliency in the face of adverse conditions. However, such studies on developmental pathways and protective factors have not been performed in children with significant biomedical as opposed to psychosocial risk, such as those with extreme prematurity. What is clear is that the future of premature children has to be looked at from a lifespan perspective, as "recovery" may not be evident until adulthood.

Myelomeningocele

Mandy was first diagnosed with an open meningo-myelocele as a fetus at 19 weeks, evaluated as moderately severe, with a lumbar lesion, anatomically at about L3, an Arnold Chiari malformation with mild ventriculomegaly, club feet, and little or no movement of the lower limbs visible. What is the QoL expectation for Mandy? Should the conversation with her parents integrate QoL considerations, and if so how?

A purely "medical" counseling session with such an expectant mother might proceed as follows: "Your fetus has a condition which can cause serious short- and long-term problems; the most immediate risk is of infection, meningitis, because the skin is not intact over the back and germs can invade. Without surgery to close the hole in the back within the first few days of life, your baby will eventually develop this life-threatening infection. Surgery will not improve the function of the legs, or the bladder or bowels, and your baby may well require another surgery soon afterward to reduce the pressure on the brain. After the surgery we will start other interventions; the most important in the short term is to teach you to catheterize your baby several times a day to drain the urine and reduce the risk of urine infections, which can affect long-term kidney function if they are frequent. Other complications are possible, complications of the surgeries, for example. In the long term most infants with this level of defect can get around on their own, perhaps with physical aids, but many will need other surgeries.

Constipation can be a major issue. In the long term many infants with this condition have some difficulty at school."⁵⁴ In contrast, a discussion which, in addition, approaches the subject of QoL would include something like: "despite all of these problems, most children like Mandy, when they are older, report a QoL which is good or very good,¹³ and most adolescents state themselves that their QoL is quite acceptable. The things which most affect long-term quality of their lives are their relationships with their friends rather than their functional limitations."⁵⁵ Although HrQoL is clearly lower than a child without any chronic health problems, there is no clear relationship between the degree of disability and QoL.⁵⁶ There are now many procedures which can improve some functional limitations, such as for example, surgeries to promote urinary continence, but these procedures probably won't affect how Mandy evaluates her QoL.⁵⁷ On the other hand provision of required resources substantially improves HrQoL.⁵⁸ In our opinion, addressing the experience of individuals who are living the consequences of medical decisions is an appropriate, indeed essential, part of the counseling process. Ignoring the adaptability and resilience of our patients is as inappropriate as ignoring the surgeries that will be required.

Hypoplastic Left Heart Syndrome

Infants with hypoplastic left heart syndrome may be diagnosed in utero as a fetus or may present after birth, as happened to Alberta, a little girl who presented because of an acute collapse on the third day of life when her patent ductus arteriosus closed.

Counseling of Alberta's parents regarding appropriate therapy should include analysis of risks in the short term, hazards of future interventions, and functional status, cardiac and noncardiac, if she survives, as well as a discussion of how patients, such as them, cope with the medical condition, and how such patients view their own QoL. For this particular condition, which until relatively recently was uniformly lethal, the medical literature includes hundreds of articles about surgical techniques and success rates of the multistage "palliative" surgeries required⁵⁹ and of the alternative of cardiac transplantation. Several articles about functional outcomes, both cardiac^{60,61} and neuro-developmental,^{62,63} and very few articles about QoL, in only one of which was an accepted QoL measurement tool used for the 18 families studied,

were found.^{62,64} This progression is entirely reasonable as the surgical approaches have evolved, but further investigation of the long-term effects on lifestyle and on HrQoL are required to adequately counsel parents.

Quality of Life and Permanent Disability

Physicians have a tendency to answer questions, such as “what kind of life will he/she have?” with a recitation of medical complications and risk assessments, likelihood of survival, and proportions of survivors with certain major complications. For some parents that is exactly what they wish to hear, but frequently a bare recital of such facts leaves out extremely important aspects of “what kind of life”: the capacity for happiness, the joy and pain of living with a child who has on-going medical problems, the satisfaction of doing a good job as a parent, or the heartache of failure.

Parents being counseled after a diagnosis of trisomy 21 frequently report later that the information was presented in a uniformly negative light, and no positive aspects of their potential future family life were discussed.^{65,66} This is true both for antenatal diagnosis among mothers who continued their pregnancy and among postnatally diagnosed families. Although for many health care providers a diagnosis of Down syndrome is seen as a disaster, parents who are carrying a fetus, or who have just delivered an infant, need and want a balanced view.⁶⁵ The positive aspects of the life that awaits them are as important as a recitation of all the potential problems.

For some conditions that are represented in this issue, we could find no QoL studies in the literature; this includes trisomy 18 and trisomy 13. Families of infants with these conditions were until recently uniformly counseled that the condition was lethal, and very few studies evaluating the functional status of the survivors have been published, and none regarding the QoL of them and their families. This has been one of the self-fulfilling prophecies of pediatrics, as such infants were thought to have a lethal condition, no intensive support was offered in the neonatal period or during later acute illness. Thus few infants survived, a 5% survival to 1 year of age often being quoted. Attitudes to such infants appear to be changing, and as a result, it appears that even with intensive care support life span is usually limited, but also quite variable, and among infants with trisomy 18 receiving

full supportive care 25% may survive the first year.⁶⁷ Many children with these trisomies are now surviving for several years: to counsel the parents we need good quality inception cohort studies of infants receiving active medical care, to describe life expectancy, functional capacities, and QoL.

Informally, from narratives, such as those in this issue, we know that parents’ experiences of living with a baby who has trisomy 18 or 13 (or other profoundly handicapping and life-shortening conditions) vary widely. Many parents adapt to the difficult life which fate has provided for them and find some sort of fulfillment and meaning in that life. We should always remember that the parents facing these issues are all individuals. There is no “typical” way of coping, and some parents find lifelong 24-hour caring for a child who has little ability to communicate and return their affection, a dependent child who may become a dependent adolescent and then a dependant adult, intolerable.

We could make a prediction, based on our understanding of the overall pattern of QoL studies among infants with chronic health conditions and their families, of what a study of a substantial number of parents of infants with trisomy 18 and 13 would look like. On certain dimensions the HrQoL of the infants would be very reduced (those regarding social skills and communication); on others (pain and contentment) they would probably perform similarly to controls. We would also conjecture that families who are able to find the social resources that they require would have children whose HrQoL is significantly improved. Families’ average scores would probably suggest that QoL is reduced compared with controls but with relatively small differences compared with families with healthy children; those average scores would hide a great deal of variability between some families who find peace, fulfillment, and a degree of contentment, and those who find pain, disappointment, and frustration (and of course those who find both, who we would guess would be the majority) in coping with such a profoundly serious diagnosis.

Conclusions

QoL evolves as life progresses. Health care decisions for children require projections of future QoL by proxies, projections which eventually become realities for the individual themselves, realities which may or may not match those projections.

As a child advances in life, physical and biomedical factors diminish in importance. In their place, psychosocial elements predominate as determinants of self-perceived QoL.

It is interesting to see that the further a child advances in life, the importance of physical and biomedical factors in determining QoL diminish in favor of psychosocial elements. Even if physical factors do not disappear from the reality of what is experienced by individuals and their families, we note that some attach less importance to them than others. Interestingly, young adults who were VLBW have a reduced objective QoL, due to the important impact of physical disabilities on this scale; subjective QoL, which is determined more by psycho-social elements, shows much fewer differences between ex-VLBW and their NBW peers.⁴³

What appears of crucial importance for people to make this transition from pure biomedical factors, such as a summation of their disabilities, to social and relational integration is probably related to the capacity of resilience.⁶⁸ Resilience is a feature of individuals, such as the ability to overcome physical disabilities and develop social and relational skills, and also a feature of families. Families often bring their experiences with a baby who is likely to be impaired by projecting suffering related to physical disabilities as the overwhelming consideration. It is only through life experience, living with a disabled child, learning how to know and support them that the family can discover positive capabilities and possibilities that open themselves throughout the child's development. As a parent of a child with serious impairments said: "The fact that the result is wonderful doesn't mean that the road has not been painful."

Projecting impairments in the neonatal period leads one to expect loss of functionality and the difficulties that will follow. In contrast, the progres-

sive adaptation to those difficulties, the resilience inherent in many families and individuals, is less immediately obvious. As noted above, paradoxically, adults with irreversible chronic conditions

have better adaptation and higher QoL than those with the same condition, but with a possibility of improvement.¹⁵ Adaptation to a chronic health condition therefore seems to require a certain degree of acceptance; hoping for improvement can paradoxically lead to being less happy. Living with a disability allows the resilient individual to concentrate himself not just on the loss of autonomy, but also on the possibilities that exist despite those disabilities.¹

Zack is celebrating his 21st birthday tonight. His best friend and his girlfriend are invited to his parents'. During dinner, they talk about Zack's project to move out with his girlfriend in a small apartment. It's a little later than all his friends who are already living on their own, but Lucy and Marc are not in a hurry to let him go by himself.⁶⁹ Zack has been working for 2 years now at

the school cafeteria and is enjoying his job. He always wanted to work with children and he's glad to be appreciated by his colleagues, even if he is still a little shy with them.

Later that night, when everyone has left, Lucy and Marc have another remembrance; they secretly think back to this terrible night 21 years ago. Lucy cries, not knowing if it is of joy or of sadness. What if they had chosen a different way? What if they had listened to their terrible fear during these few hours when their destiny could have been completely different? Life has been difficult since then, but definitely

not as bad as they feared. Zack is a happy young man who is enjoying his existence, what could be better for a parent?

Life has certainly not been as they had planned; is it ever?

As a child advances in life, physical and biomedical factors diminish in importance. In their place, psychosocial elements in predominate as determinants of self-perceived QoL.

Hoping for improvement can paradoxically lead to being less happy. Living with a disability allows the resilient individual to concentrate himself not just on the loss of autonomy, but also on the possibilities that exist despite those disabilities.

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