The history of childhood disabilities

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Abstract

Appreciation of the uniqueness of childhood can foster awareness and recognition of the special needs for children. The aims of this article are to delineate the definitions of childhood disability and describe childhood disabilities through the ages with a chronological perspective and a focus on dental disabilities. Patients with the disabilities associated with the Ectodermal Dysplasias and Naevoid Basal Cell Carcinoma Syndrome are used to illustrate the effects on oral health.

Key words: Disability, ectodermal dysplasia, naevoid basal cell carcinoma syndrome, social construction

Introduction

The destiny of children with physical impairments was long dependent on their families’ circumstances. Historical study of the issues relating to children and disabilities will yield a better understanding of how societies have treated these individuals over time and in the present (Safford, 1996; Schalick, 2001). Appreciation of the uniqueness of childhood can foster awareness and recognition of the special needs of children. This article will delineate the definitions of childhood disability and describe childhood disabilities through the ages with a chronological perspective and a focus on dental and medical disabilities. Three predominant themes will be examined illustrating how:

• Childhood disabilities have varied medically, culturally and socially over time
• A social campaign addressing the needs of disabled children, in the setting of modernity, was affected by biomedical approaches around disability
• Social attitudes can influence the medical model.

These themes will be substantiated in the context of discussion of the trends in infant mortality, the influence of the Progressive Movement, and will contextualise experiences with the child with physical disabilities, as the central theme, using primary and supportive secondary sources. Conditions with unique significance to the history of disabled children will be scrutinised, and how certain conditions, conceived as biomedically redressed, were superseded by others.

Definitions of childhood disabilities

Defining and measuring disability in childhood has been a challenging task. Infancy to adolescence is a period marked by dynamic changes in the body structures and functions; the acquisition of physical and mental skills; and progressive steps toward independence of movement, thought, and behaviour (Simeonsson, 2006). The evolving characteristics of the child complicate the task of assessing function and distinguishing significant limitations from variations in the normal developmental processes. The younger and less mature the child is, the greater the challenge has been to define disability. Infancy (0-2 years) simulated disability when limbs are inadequate for walking and when there is no articulate speech. This is in contrast to the relatively stable characteristics of the adult. In the history of childhood disability, earlier approaches to defining disability took the form of observing physical signs or noting discrepancies in the appearance of basic maturational skills. Disability in childhood and adulthood is intrinsic to the human condition where there will always be some people with disabilities.

In the USA, the Individuals with Disability Act (IDEA), renamed in 1990 from the 1975 The Education for All Handicapped Children Act (EAHCA), defines a “child with a disability” as a child with neurodevelopmental delay, hearing impairments (including deafness), speech or language impairments, visual impairments (including blindness), serious emotional disturbances, orthopaedic impairments, autism, traumatic brain injuries, other health impairments, or specific learning disabilities, and who [because of the condition] needs special education and required services. This law ensures services to children with disabilities throughout the United States.

IDEA governs how states and public agencies provide early intervention, special education and related services to more than 6.5 million eligible infants, toddlers, children and youths with disabilities. Children with disabilities who qualify for special education are also automatically protected by the Rehabilitation Act of 1973 and the Americans with Disabilities Act (ADA).

The 1990 Americans with Disabilities Act (Americans with Disabilities Act of 1990, 1990) left room for inter-
pretation of the definition of disability: The term “disability means, with respect to the individual: (A) a physical or mental impairment that substantially limits one or more of the major life activities of such individual; (B) a record of such an impairment; or (C) being regarded as having an impairment.” There is much subjectivity in this definition and that evaluation of an individual claiming disability is on a case-by-case basis. The WHO (World Health Organisation, 2001) defined impairment as any loss or abnormality of psychological, or anatomical structure or function; disability as any restriction or lack (resulting from an impairment) of ability to perform an activity in the manner or within the range considered normal for a human being; and handicap as a function of the relationship between disabled persons and the environment and occurs when there are cultural, physical or social barriers encountered which limits their opportunities and prevents access to the various systems of society that are available to other citizens.

The way disability is defined and understood has changed in the last decade. Now, the WHO has moved to a new international classification system, the International Classification of Functioning, Disability and Health (ICF) (International Classification of Impairment, Disability and Handicap, 2001) which emphasises functional status over diagnoses. The ICF also calls for the elimination of distinction between health conditions that are ‘mental’ or ‘physical’ and focuses on analysing the relationship between capacity and performance. If capacity is greater than performance, then the gap should be addressed through both removing barriers and identifying facilitators. The WHO now defines disability as “a contextual variable, dynamic over time and in relation to circumstances”. The ICF also acknowledges that the prevalence of disability is influenced by social and economic environments.

The US Senate Finance Committee in 1995, approved legislation creating a childhood-disability definition for the Supplemental Security Income (SSI) program (American Academy of Pediatrics, 1995): a child younger than 18 is considered disabled if a child has a “medically determinable physical or developmental impairment, which results in a marked, pervasive and severe disability, and is expected to last for a continuous period of 12 months or result in death.”

Although there is no universal standard for defining disability, the ICF definition is preferred, for which there is a children and youth version, as it has benefits for both the client and the health professional. A major advantage for the client is the integration of the medical and social aspects of his or her health condition. There is benefit to the health professional because identifying the limitations of function is often the information used to plan and implement interventions such as treatment and rehabilitation. The health care professional does not have to rely solely on a diagnosis which reveals little about one’s functional abilities.

Disability in childhood throughout history

The nomenclature for children with disabilities has changed throughout history. Usage of terms such as monsters, deformed, deviant and crippled were transformed into terms like disabled, different, special needs and exceptionalities. Individuals with disabilities can be seen as both similar and yet significantly different from culture-to-culture and from time-to-time (Schalick, 2001). For children with disabilities, history has not followed a uniformly positive trajectory, despite the apparent evidence of progress (Safford, 2006). Childhood infectious diseases that once caused permanent deafness, blindness, other physical or mental impairments, have been virtually contained, at least in the United States and other industrialised nations. Meningococcal septicaemia in the infant and young child is an example of such an infectious disease that results in profound disabilities; permanent deafness, motor impairment and amputation due to peripheral ischemia and gangrene may result. However, there is a current epidemic, formerly believed to be rare, of autism spectrum disorder. Fortunately, children in the United States are no longer routinely exposed to maiming or toxic conditions in mines, mills and the marketplace. Yet, American children regularly incur permanent physical, mental, and emotional damage as a result of being shaken, sexually brutalised, or exposed to prenatal toxic influences in the form of drugs or alcohol, or to developmental toxic influences in the form of exposure to violence (Safford and Safford, 2006). It will be demonstrated with a chronological perspective, how childhood disabilities varied medically, culturally and socially, from prehistory to antiquity to Early Christianity/Medieval/Renaissance to the early modern period, 18th century Enlightenment and 19th and 20th century modern periods so that we may learn from past errors and avoid repeating them. The history of childhood disability or ‘difference’ reflects the way people and society throughout centuries, have understood, interpreted and treated ‘difference’.

Childhood disability in prehistory

The timeline of human prehistory began in the Paleolithic period about 120,000 to 150,000 years ago with the appearance of Modern Homo Sapiens and ended in 3700 BC when cuneiform writing appeared and records were kept. Prehistoric people relied on religious beliefs and practical treatments made from local materials to treat their ailments. In prehistory, physical and behavioural deviance was attributed to spirits animating the natural world, their powers invoked for protection and cure, their mischief counteracted by exorcism and trephination (Safford and Safford, 2006).
Childhood disabilities in the Paleolithic period (paleodisability) clearly existed in prehistory although the evidence is scant with no written record (Schalick, 2006). Since survival of a condition is required for the formation of skeletal lesions or abnormalities, all known cases are by definition, suggestive of some degree of survival of non-lethal disabilities (Trinkaus, 2005). Skull fragments were discovered in the Jutland peninsula of Denmark that indicated trepanation had been performed, probably by a medicine man, on a pre-historic hydrocephalic infant to remove the evil spirits. An archaeological report from Indiana, USA, presented an analysis of an adult male that exhibited diagnostic markers of cleft palate (Schalick, 2006). The burial was associated with the Grider Site, Late Woodland context, located in Pike County, Indiana, USA. This analysis demonstrated that a prehistoric culture could overcome the health issues experienced by an infant with a cleft palate and survive to adulthood.

**Childhood disability in antiquity**

In Egypt, The Roman Empire and Ancient Greek times, there was a medical interest and utilitarian use of childhood disability; for example, ‘crippled’ children were trained and even mutilated to become beggars. In antiquity, people knew of neonatal mortality and about the vulnerability of the neonate. Aristotle stated “Most of the babies are carried off before the 7th day; that is why they give the child its name then, as they have more confidence by that time of its survival” (Dunn, 2006). Aristotle had other philosophical thoughts about children with deformities and birth. He is quoted as saying “As to exposure of children, let there be a law that no deformed child shall live. However, let no child be exposed because excess populations, but when couples have too many children, let abortions be procured before sense and life have begun.” He further stated “This is the reason why mothers are more devoted to their children than fathers; it is that they suffer more in giving them birth and are more certain that they are their own.” For Greeks and Romans, monstrous births were a portent like comets, earthquakes and wild storms.

In Sparta and Athens, child-rearing was explicitly in the service of the state, in the former to prepare for war, in the latter for peace. Under the laws of Lycurgus, each newborn was brought to a panel of elders and if judged strong was entrusted to the state for military training, which began in infancy; those found unfit were exposed. In Athens, cultural rather than military training was the focus and a newborn’s fate was determined by the father, to live or to be disposed of “in clay vessels…left by the wayside” (Safford and Safford, 1996). This was actually a form of abandonment, with the implied hope that some stranger might care for the baby. This paternal decision to abandon a newborn had to be made during the first eight days, and since some impairment would not have been apparent, we can assume that many exceptional children survived. While historians have conflated child abandonment and infanticide, the former was often the alternative to the latter. The overwhelming belief in the ancient world was that abandoned children were picked up and reared by someone else. Some babies with congenital anomalies survived in ancient Greece whether by accident or design. Perhaps, passive infanticide was practiced whereby the infant would not be killed outright, but would not have received the same attention necessary for survival (Rose, 2003).

Archaeological evidence reveals instances of relatively long life, group inclusion of members with physical impairments, and even attempts at surgical correction. After all, a deaf child of a farmer could still work in the fields. Therefore, the criteria for physical disability relied on one’s functional ability within the community (Community Model of Disability) rather than on one’s ability to function as an individual. The application of sophisticated medical knowledge was probably minimal to children with disabilities, but this did not imply lack of concern for children or for exceptionality (Rose, 2003).

In antiquity, there was a distinction between malformation, neurodevelopmental delay and illness (Stiker, 1997). The blind, deaf and feebleminded were not categorised among the deformed and public resources were made available to those citizens who were denied a living by disability. Hippocrates (Greece, 460 BC-370 BC) is credited with being the first physician to reject superstitions, legends and beliefs that credited supernatural or divine forces with causing illness (Lloyd, 1983).

**Childhood disability in early Christianity and the Middle Ages**

During early Christianity and the Middle Ages (which began with the destruction of the empire in 476), the disabled child was sometimes viewed as a reflection of their parents’ sins and was based upon religious stereotypes and isolation. Medieval societies did, however, identify exceptionalities as being undesirable but were integrated into the community since archaic societies could not afford to support unproductive members. In *The Confessions* of St. Augustine (356-430) he reflected on his own memories of childhood, attributed them to the result of sin, inherent propensities for evil, and children had to be redeemed. This doctrine of original sin had profound implications for children with disabilities, and for their parents; these children represented punishment for parental sin or the working of Satan. Children generally had a pre-
disability to children was recognised (Grant, 1974). The following statement was made at the time: “It is known by everyone that we ought not to do violent surgical operations on small children, as we do on young men, adult and old people, such as phlebotomy, cautery, incision, nor give theriaca or a laxative” (Bracton, 1220). It was also stated that “small children and decrepit old people need smaller doses of medicines and are in grave danger from poisons (especially less than 10 years-old).” Another statement by Henry Bracton in the 1280s, reflects some of the beliefs of the time that “Any child which is born a monster-as one that has more than the proper number of members…or a deficiency of the same…such children shall not be administered to any inheritance, accounted not as children but as beasts or monsters.” By 1500 there was unfortunately evidence that parents or guardians of monstrous children took them from town to town to show for money; some cities even began to issue licences/permits for spectacles of this sort (Bracton, 1220).

Childhood disability in the early modern period

The early modern period refers to the period roughly from 1500 to 1800 in Western Europe and was characterised by the rise of science, improvements in transportation and communications, increased technological progress and secularised civic politics as well as state intervention.

The Early Modern period and a portion of the Enlightenment was the transition point away from the medieval model of Church authority and feudal relationships at the community level into a state, centralised, and bureaucratic set of relationships that could place people with disabilities into a box or category, labelled as ‘not able to work’, a system that frustrated the disabled (Schalick, 2009a). The Poor Act (1601) formalised earlier practices for poor relief. The Act made provision to: board out young orphaned or not cared-for children, with payments to families willing to accept them, provide materials to “set the poor to work”, offer relief of people who were unable to work (lame, impotent, old and blind), and “the putting out of children to be apprentices.”

Up to the 1700s, society in general did not offer to children with special needs the possibilities of formal education. This care and protection of children with special needs appeared only in the 18th century.

Childhood disability in the 18th Century

The Enlightenment is a term that describes a time in Western philosophy and cultural life centred in the 18th (1650-1800 or 1700-1800) century, in which reason was advocated as the primary source and legitimacy for authority; it was during this time that the philosophers attempted to understand and develop the needs of children. This period was driven by reason and there was a social contract that supported the idea that legitimate state authority must be derived from the consent of the gov-
erned. The disabled were often left out of this implicit contract (Schalick, 2009b). Jean-Jacque Rousseau’s, The Social Contract in 1762 outlined a social contract based on popular sovereignty which meant that people give up some rights to a government to receive social order but that legitimate state authority must be derived from the consent of the governed. He also insisted on children’s rights to be happy and develop their own natural gifts. The Enlightenment led to core ideas such as freedom, democracy, the scientific method, religious tolerance and a concept that social/environmental modification could improve humans and society. This period also witnessed Residential Schools for the Deaf and Blind that were established in 1755 in Paris. Prior to 1700, doctors were rarely at childbirth, but were rather attended by family, friends and neighbours.

**Childhood disability in the 19th and 20th Centuries**

Modernism (1884-1914) affirms the power of human beings to create, improve, and reshape their environment with the aid of scientific knowledge, technology and practical experimentation but again, the disabled were often left out of the implicit contract (Schalick, 2009c). Galton believed that late marriages of eminent people and the paucity of their children were dysgenic and encouraged eugenic marriages by supplying incentives for those able to have children.

World War I produced unique historical and representational convergences between children with physical disabilities and wounded soldiers in Great Britain (Koven, 1994). Koven, in his historical investigation, traced the intermingled roles of children with physical disabilities and disabled veterans returning from World War I in Great Britain where there were social challenges created by the War, which forced these two groups into an odd relationship. The veterans were infantilised by society, while the children were glorified as ‘little soldiers’. Both demonstrated how social conditions from poverty to poor education created a ‘crippling’ culture and suggested mechanisms for change.

Children in Nazi Germany who failed to comply with the social or biological criteria of ‘perfect’ children were taken from their homes and communities, isolated in institutions, hospitals, work and concentration camps, or murdered (Rowgow, 1998). These unwanted children were comprised of orphans, children with emotional or behavioural problems as well as children with mental and physical disabilities. Most reforms implemented during the Weimar Republic were abolished, including provisions for educational opportunities for disadvantaged and disabled children. Pre-school programmes for blind or deaf children were eliminated.

In the 1960s and 1970s, the Scandinavian countries, along with Italy, were leaders in regard to integration into society of people with disabilities (Solvang et al., 1993) In the 1950s and 1960s, there were four groups of disabilities identified which were defined on the basis of their medical diagnosis, and included the blind and visually disabled, the deaf and impaired, the intellectually disabled, and the mobility disabled. The policy documents concerning the normalisation reform viewed these different disability groups as one. In the mid 1960s, about half of the estimated population of severely disabled children in Sweden was segregated into institutionalised education. In Norway and Sweden, there evolved integration of disabled persons into conventional schools where the disabled person was not be viewed as handicapped in her- or himself, but the handicap was shaped by society, physically by architectonical barriers and socially by prejudices towards people with disabilities.

After the collapse of Communism in 1990 and the newly-established democracy, the massive mistreatment of abandoned, sick, and disabled children housed in Romanian institutions, was revealed (U.S. Embassy, 2001). Due to the rampant national poverty of the preceding two decades, there was institutionalisation of more than 170,000 children since there were no community-based childcare alternatives or civil society involvement. Between 1996 and 2001, there were attempts at dismantling the warehousing of children in Romania. The solution also involved the building of family-oriented, community-based services for children, based on child protective service models in Western Europe and the United States.

Accelerating forces of nationalisation and state bureaucratisation placed people with disabilities in narrower boxes or categories in the 19th and early to mid-20th century. These categories revolved around labour, labels, state support and remedial assistance in an increasingly technological and literate environment. The features of ‘modernity’ isolated people with disabilities. The medical model later became more refined at this time while balancing medical intervention and social integration; the social model became the basis for a civil rights/justice oriented effort in the second half of the 20th century (Schalick, 2009c). There was a changing and competing impact of bioscience on the meaning of children with disabilities in the 20th Century. Furthermore, parents became more included in the care of the disabled child and there was more therapeutic optimism.

Today, more than half of the incidence of childhood disability worldwide results from malnutrition, viral and bacterial infections and communicable disease. In America, one child in four is born into poverty who becomes predisposed to disability due to manifesting a disproportional consequence of organic diseases and accidents. Many go without immunisation, are affected by violence, have prenatal exposure to drugs or toxic environmental elements, are exposed to HIV-AIDS, all created by poverty and society.
In the following sections, it will be examined how a social campaign addressing the needs of disabled children, in the setting of modernity, was affected by biomedical approaches around disability and will illustrate how social attitudes can influence the medical model. Infant mortality, the Progressive Movement, clinical experiences, and conditions with unique significance to the history of disabled children will be exemplified to illustrate these points. In the modern era, there was a trend of thought that affirms the power of human beings to create, improve, and reshape their environment with the aid of scientific knowledge, technology and practical experimentation. This period experienced medical narrowing, medical specialisation, medical insurance, asepsis, and anaesthesia which influenced the treatment of disabled children.

The progressive movement and the child with physical disabilities

The Progressive Era in the United States was a period of reform which lasted from the 1890s to the 1920s. Typically, progressive reformers exhibited a strong sense of social justice that led to the aid of many marginalised groups in American society. Although the progressives failed to fully appreciate the complexity of the problem with disability, they did realise there were physical and cultural impediments and barriers between disabled children and the public school system. A loose coalition of socially conscious reformers, as part of the progressive movement, promoted a movement to aid disabled children (Byrom, 2006). The author, Brad Byrom, demonstrated how the progressive emphasis on science and education affected the new charitable and public policies regarding disabled children. Instead of designing reforms that addressed the essential problems facing disabled children, reformers created institutions that reflected “their affinity for science and the expert”. They created institutions in which experts could apply their knowledge; yet in the process, they literally and symbolically, further separated disabled children from their non-disabled peers. This resulted in a new and evolving definition of disability: “it was not the physical impairment that caused problems for the disabled individual, but society’s treatment of the individual.” This language mirrors that of the more modern “social model” of disability that is at the core of the disability rights movement. American Magazine, a periodical publication founded in 1906, published an article by Gray in 1922 (Gray, 1922), entitled “Joe Sullivan’s Body is Weak but not his Will”. Joseph F. Sullivan, who experienced firsthand the difficulties facing ‘cripples’ and later became an ardent proponent of education for disabled children, demonstrated the frustration that many disabled people felt concerning the matter. Sullivan displayed both intellectual curiosity and stubbornness in his pursuit of knowledge. He had lost the use of his legs and one arm following the onset of poliomyelitis at age four. He felt ‘the sting’ of a label that begrudged him from a ‘normal’ childhood and wrote “almost everyone you met, stared at you.” This exemplifies a social attitude that impeded many disabled individuals from reaching their full potential despite medical advances.

Prevention of infant mortality

Biomedical advances and initiatives by organisations such as the Centers for Disease Control have led to slightly declining infant mortality in the United States; yet 28,000 children under the age of one still die every year (Bakalar, 2009). “The main reason for this high rate is preterm delivery, and there was a 10% increase in such births from 2000 to 2006,” according to the figures from the Centers for Disease Control and Prevention. In 2009, the latest year for which world data are available, the United States had a higher infant mortality rate than 28 countries, including Singapore, Japan, Cuba, and Hungary. In 1960, the United States had a higher rate than only 11 countries. Thus, the US is still struggling with infant mortality trends. There are large differences by race and ethnicity. Non-Hispanic black, American Indian, Alaska native, and Puerto Ricans have the highest rates of infant mortality. These figures are a concern for researchers and policy makers to make appropriate adjustments and avoid indifferent or biased social attitudes. In an article published by the Centers for Disease Control in 2008 (Mcdorman and Matthews, 2008), it was indicated that “infant mortality is one of the most important indicators of the health, quality and access to medical care, socio-economic conditions and public health practices.”

Infant mortality is defined as the number of deaths of infants (one year of age or younger) per 1,000 live births (MMWR Weekly, 2004). The most common cause of infant mortality worldwide has traditionally been dehydration from diarrhoea, but due to educational dissemination, this cause of infant mortality is now second to that of pneumonia. Major causes of infant mortality in more developed countries include congenital malformations, infections and SIDS. Neonatal mortality only includes deaths in the first 28 days of life and child mortality includes deaths within the first five years after birth. Infant mortality correlates very strongly with, and is among the best predictors of, state failure. For example, one of the highest rates of infant mortality is in Angola which has 180.21 deaths per 1,000 live births. The US Department of Health and Human Services reported that infant and neonatal mortality rates demonstrate racial/ethnic disparities (US Department of Health and Human Services, 2005).

In Richard Meckel’s Save the Babies: American Public Health Reform and the Prevention of Infant Mortality (1850-1929, 1990), it was recognised that infant survival
and health were intimately connected to the physical condition and behaviour of women during gestation, the neonatal period and infancy. Physicians throughout much of the latter half of the 19th century regarded gestation essentially as a natural physiologic process that required patience more than direct supervision and intervention. Formal obstetric involvement in prenatal care did not seriously begin until the turn of the twentieth century which contributed to the reduction of infant mortality. The availability of maternity insurance also had a favourable impact on reducing infant mortality.

Conditions with unique significance to the history of disabled children

Physical impairments in children are numerous and diverse but certain conditions, however, have had unique significance in the history of services for disabled children; epilepsy, cerebral palsy, tuberculosis and poliomyelitis.

Epilepsy, in past history, linked science and superstition, retardation and psychic disturbance. However, advances in medical science have determined the scientific basis for this convulsive disorder. The list of famous historical figures with epilepsy is impressive and includes Socrates, Alexander the Great, Napoleon Bonaparte, Vincent Van Gogh, and Alfred Nobel (Safford and Safford, 1996). Institutional confinement was endorsed for the epileptic patient who was excluded from educational opportunities. Medications for seizure control advanced from Locock’s use of bromides in 1857, to Hauptmann’s introduction of Phenobarbital in 1912, to the first non-sedative medication, diphenylhydantoin, developed by Putnam and Merritt in 1937. By the 1960s, a variety of anticonvulsant medications were differentially prescribed based on the individual’s unique situation and people became enabled to function in society.

Prenatal and perinatal causes of birth defects associated with cerebral palsy had been identified by W.J. Little. Historically, provisions for children were made in response to public health concerns and to disease rather than neuromotor or orthopaedic impairments.

Children with cerebral palsy have constituted the largest single group of children with physical disabilities identified as eligible for special education, though many do not need or receive special education (Safford and Safford, 1996). Mobility and communication needs in children; epilepsy, cerebral palsy, tuberculosis and poliomyelitis.

Tuberculosis is a disease that can be traced throughout history and experienced resurgence in the form of drug-resistant strains early in the 1990s after public health measures in industrialised nations had seemed to significantly control its prevalence and impact (Safford and Safford, 1996). Most children’s hospitals in the late 1800s would not admit children with contagious diseases or ‘chronic incurables’. However, in 1904, a model emerged that fostered open-air facilities used for education and treatment. By the middle of the twentieth century, tuberculosis was no longer the focus of specialised schooling and was not thought to be a major public health problem in developed nations.

A model emerged in the 1940s in response to poliomyelitis, known as ‘infantile paralysis,’ a condition that Americans knew had affected their president’s mobility, though few knew how much (Safford and Safford, 1996). The youth of today do not appreciate the impact that poliomyelitis had on families before the contributions of Salk and Sabin.

In addition to epilepsy, cerebral palsy, tuberculosis and poliomyelitis, there are additional disabilities in children that dominate today’s society: childhood deafness, blindness, cognitive disability, intellectual retardation, and other physical disabilities but there have been attempts through legislative enactments, to assist these disabled children with varying degrees of success.

Disabled children with an Ectodermal Dysplasia and Naevoid Basal Cell Carcinoma syndrome

The 2000 definition by the American Academy of Pediatric Dentistry of dental disability states that a person should be considered to have a dental disability if pain, infection, or lack of functional dentition which:

1. Restricts nutritional intake adequate for growth and energy needs
2. Delays or otherwise alters growth and development, or
3. Inhibits participation in life activities.

Birth defects affect 5% of all infants born in the United States, and three-quarters of these involve the head, face, and oral tissues. Two genetic disorders illustrate the place of dental anomalies in childhood disabilities.

Ectodermal Dysplasias

This first case illustrated is a patient that has been treated and followed over 20 years and is one of the X-linked hypohidrotic ectodermal dysplasias (Christ-Siemans-Touraine Syndrome) and published in 1993 by Smith and Vargavirk (Smith et al., 1993). The proband (male) exhibited the complete phenotype of CST-ED. His mother, who is the carrier of the trait, demonstrated some of the clinical features (hypotrichosis, hypodontia, and mild hypohidrosis) and two female siblings, also carriers, exhibit-
limited mild features of the disease. At the age of 5 years, the boy was referred for evaluation for reconstruction with osseointegrated implants. Clinical evaluation revealed a hyperactive child with dry hypo-pigmented skin. The hair was thin, sparse, and blonde with sparse eyelashes and eyebrows. There was congenital absence of anterior and posterior teeth in the maxilla and mandible in both the primary and permanent dentitions (Figure 1). The anterior teeth that were present were cone-shaped.

The 5-year-old child was teased because his appearance was different and because he was missing most of his teeth which created an urgent need for tooth replacement. To construct a prosthesis with stability and retention in the lower arch, it was elected to place one 13-mm osseointegrated implant in the anterior mandible, which was the only potential recipient site available (Figure 2). The implant became osseointegrated and a removable prosthesis was constructed with an O-ring attachment (Figures 3 and 4). The patient has been followed for over 20 years and the implant is still present and functional.

This patient had several supportive elements in managing this disability. The family was very supportive, particularly the mother. There was multidisciplinary care available in the Center for Craniofacial Anomalies, located at the University of California, San Francisco, which has been caring for individuals with craniofacial differences since 1954. This exemplifies the concept of the dental home, derived from the definition of the American Academy of Pediatrics of a medical home, which states paediatric primary health care is best delivered where comprehensive, continuously accessible, family-centred, coordinated, compassionate, and culturally-effective care is available and delivered or supervised by qualified child health specialists. State support was provided by the California Children Services. National organisations such as the National Foundation for Ectodermal Dysplasia and the Ectodermal Dysplasia Society provided patients and parents with information regarding genetic counselling and testing. This is an example of a patient that the dental profession in the past may have been referred to as a ‘dental cripple’ because of his hypodontia and masticatory dysfunction. The medical and social support for this child balanced medical intervention with social integration enabling him to have improved function and appearance, attend school with reduced harassment and develop into a productive young adult, integrated into society.

The earliest recorded cases of ED were described in 1792 (Yavuz et al., 2008). This disorder is considered relatively rare, with a rate of 1 in 10,000 to 1 in 100,000 births. In its fully manifest form, hypohydrotic ectodermal dysplasia (HED) leads to typical dysmorphia of the face, referred to as ‘old man’ facies. Adaptive functioning of these individuals is dependent on the severity of the symptoms. The child’s intellectual potential and personality, how the disease is dealt with within the family, and reactions from the child’s environment influence adaptive functioning in different ways (Hummel and Gudda, 1997). Affected patients mostly feel handicapped because of their facial and oral appearance. These children are often excluded by their peer group, as was the situation in this case, and become the target of criticism and mockery. The intelligence level of the afflicted children, a history of somatic, psychosomatic, and psychosocial problems, as well as the reactions of the patients’ peers and adults within their immediate social environment forms a complex, intertwined network that ultimately determines the individual’s adaptive behaviour. (Hummel and Gudda, 1997).

The psychosocial stress and adaptive functioning of such patients was investigated by means of a semi-structured interview with questionnaires, conducted on a sample of 14 children and adolescents with varying degrees of this condition (Hummel and Gudda, 1997). The results revealed that adaptive functioning is not only dependent on the severity of symptoms but also on the intellectual potential and personality, how the disease was dealt with within the family, and reactions from the child’s environment.

**Naevoid Basal Cell Carcinoma syndrome**

This case of Nevoid Basal Cell Carcinoma syndrome (NBCCS) or Gorlin-Goltz syndrome is in a patient who has been followed for more than 20 years. This condition is an autosomal-dominant inherited disease, exhibiting high penetrance and variable expression (Leobardi et al., 2009). The prevalence is estimated at 1 in 56,000. NBCCS is characterised by the development of multiple basal cell carcinomas of the skin, odontogenic keratocysts of the jaws, palmar and plantar pits and abnormalities of the face (frontal bossing, hypertelorism). Other less frequently seen characteristics are spine and rib abnormalities and calcification of the falx cerebri. It is also associated with other malignancies.

The patient was a 14-year-old girl who was referred to the Oral and Maxillofacial Surgery Department, at the University of California, San Francisco, USA. She presented with hypertelorism, a short fourth metacarpal, basal cell carcinomas of the skin (Figure 5), and multiple odontogenic keratocysts of the maxilla and mandible (displacing the developing third molars) (Figure 6). The odontogenic keratocysts and the associated impacted teeth were removed under general anaesthesia (Figure 7). Other finding included calcification of the falx cerebri, pectus excavatum, bifid ribs, and bifid spine. The best known orofacial manifestations of NBCCS are frontal and parietal bossing, increased occipitofrontal circumference, mild mandibular prognathism, and mandibular coronoid processes hyperplasia. Nevoid basal cell carcinoma syndrome left evidence of its existence in mumified Egyptian skeletons nearly 4000 years ago, but was
not described until much later when it was delineated by Gorlin and Goltz in 1960 and bears the former’s name (Ratner, 2004).

The patient was offered genetic counselling as part of her management. The BCCNS Support network was available to provide healthcare, counselling and support services to children and adults with this condition. She did develop ovarian fibromata, which can be part of the syndrome. She had children who all manifested characteristics of nevoid basal cell carcinoma syndrome that also required treatment.

Figure 1. Partial anodontia with malformed dentition in a child with ectodermal dysplasia

Figure 2. Osseointegrated implant placed in the mandibular midline

Figure 3. Prosthesis with “O” ring to retain prosthesis

Figure 4. Prosthesis in place in the mandible

Figure 5. Basal cell carcinoma of skin

Figure 6. Panoramic radiograph showing cystic lesions in all four quadrants and displaced 3rd molars

Figure 7. Excised odontogenic keratocyst enveloping impacted 3rd molar

Table 1. Infant Mortality Rates 2009 World Comparison (Central Intelligence Agency USA, The World Factbook)

<table>
<thead>
<tr>
<th>Country</th>
<th>Deaths/1,000</th>
<th>Country</th>
<th>Deaths/1,000</th>
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<th>Deaths/1,000</th>
<th>Country</th>
<th>Deaths/1,000</th>
</tr>
</thead>
<tbody>
<tr>
<td>Angola</td>
<td>180.21</td>
<td>India</td>
<td>30.15</td>
<td>United States</td>
<td>6.26</td>
<td>Singapore</td>
<td>2.31</td>
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<tr>
<td>Sudan</td>
<td>82.43</td>
<td>China</td>
<td>20.25</td>
<td>European Union</td>
<td>5.72</td>
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</table>
Discussion, synthesis and conclusions

This paper offers a synthesis of themes and topics relevant to the history of childhood dental and other disabilities. The relative absence of children in standardised history texts has obscured children in history. There are, however, images of disabled children, found throughout well-known literature, film and even opera, with their characters ranging from sweet, brave and tragic. Disabled children also served as ‘poster girls and boys’ for fundraising drives and causes. These images reflected a historical presence established by the society in which the children lived. Historians are limited to the evidence of the past that has survived to the present, as well as to primary and secondary sources which vary widely depending on the time and place.

The opportunities and treatment of children with special needs from an historical perspective were formed by the attitude towards disabled children dictated by each period of history, and by the depth of understanding of the children with exceptional and special needs. This understanding is deeply rooted in religious, ethical, medical, political, and economic aspects, as well as society’s attitudes towards what is the norm. Integration and inclusion of these disabled children, rather than isolation and segregation, has been and will continue to be dictated by public attitudes and medical judgments.

On the progress side, infant mortality has been significantly reduced, responsible social legislation has been enacted, socially conscious organisations have emerged, and conditions with unique significance to children have been diminished in frequency and intensity; yet disparities in children’s oral and general health, as well as access to dental and medical care, still persists today. The two cases, presented in this paper, illustrate how a modern and more refined model meant that medical intervention and social integration were more balanced, and enabled these two patients, with potentially significant disabling conditions, to avoid segregation and isolation. They were able to benefit from integration and inclusion and to experience their fullest human potential. These two individuals had disorders with resultant impairments, but with limited disabilities and minimal handicaps due to medical technology and social services available to them.

When the earliest recorded case of ED was reported in 1792, there was no genetic counselling, no understanding that these individuals were prone to childhood febrile respiratory illness that may result in reduced intellectual development, no computed tomography and no prosthodontic capabilities and implant technology to remedy the functional impairments and the aesthetic images. This disease was identified during the 18th century period of Enlightenment when philosophers attempted to understand and develop the needs of children but it was unlikely that any of the children with this disorder, received any significant treatment to prevent a more profound disability and handicap, and probably associated ridicule.

There were numerous supportive elements in managing this disability including the National Organization for Rare Disease (NORD/Ectodermal Dysplasia (National Organization for Rare Disorders, 2007) and the Ectodermal Dysplasia Society (Ectodermal Dysplasia Society, 1984) which provides patients and parents with information regarding supportive services, genetic counselling and testing.

The Neviod Basal Cell Carcinoma Syndrome was delineated by Gorlin and Goltz in the 1960s but most certainly was observed in previous historical times because of the external manifestations. Although surgical and rehabilitation services were readily available in the 1960s, more sophisticated techniques and imaging only became available later, which improved outcomes enabling enhanced social integration.

There is a Basal Cell Carcinoma Nevus Syndrome Support Network (Network of Care, 2010) to provide healthcare, counselling, and support to children with genetic disorders. They also develop information about this medical condition by those living with the disorder and share information with clinics and health care providers treating this entity. More importantly, the organisation provides a supportive environment for families to seek treatment for their children and train healthcare providers to communicate more effectively with families. An online forum is available to see what the concerns are of the members. These organisations work very firmly with the medical model of disability since the children have conditions which need to be addressed medically, not just socially.

Both these genetic disorders have phenotypic manifestations that can lead to labels and stigma. The Network of Care for Behavioral Health (Network of Care, 2010) educates individuals and families with genetic diseases and describes how stigma discourages people from getting help, keeps people from getting good jobs and advancing in the workplace. Stigma is a barrier and discourages individuals and their families from getting the help they need due to the fear of being discriminated against.

Childhood disabilities varied medically, culturally and socially from prehistory to Early Christianity/Medieval/Renaissance to the early modern period, the 18th Century Enlightenment and 19th and 20th century periods which gives us the opportunity to learn from the past and transform the future. The history of childhood disability or ‘difference’ reflects the way people and society throughout the centuries have understood, interpreted and treated ‘difference’. Individuals with disabilities can be seen as both similar, and yet significantly different, from culture-to-culture and from time-to-time.

In prehistory, physical and behavioural deviance was
attributed to spirits animating the natural world which were counteracted by exorcism and trephination. In Egypt, the Roman Empire and Ancient Greece, there was medical interest in disability, and a utilitarian use of the difference, whereby crippled children were trained to become beggars. In early Christianity and the Middle Ages, difference was the reflection of their parents’ sin based on religious stereotypes and isolation. The Renaissance saw the human being as the highest value and that it was possible to educate a child with special needs Girolamo Cardano (1501-1576) (Schein and Stewart, 1995). He insisted that the deaf could learn to communicate by reading and writing, in contradistinction to Aristotle who believed deaf persons could not be educated. Cardano was one of the first to state that deaf people could learn without learning how to speak. The ideas of John Locke (1632-1704) viewed the child as a blank sheet of paper that can be filled in and offered new hopes for children with special needs. The ideas of French Enlightenment philosophers proposed to understand and to develop the child’s needs. They offered education in a separated environment, separately from the children of the comprehensive schools which have remained from the 18th century to the present. During this time, the industrialists discovered that children, especially poor ones, constituted the world’s most inexpensive labour force.

Where we stand today on childhood disabilities

A new interest about children in times past was generated by the progressive era agenda which sought to educate, acculturate, and elevate American children through child study and child welfare. The Century of the Child with the Individuals with Disabilities Education Act, the Americans with Disabilities Act, and the recognition how barriers imposed by others, contributed to the handicapped state, suggested how radically the context of exceptionality has changed. However, even as barriers are removed, children remain inevitably disabled by their very dependence and inability to choose the circumstances of their birth. In 2009, the University of California celebrated the week of the young child to recognise the needs of young children and disabilities.

Childhood diseases, such as Congenital Rubella Syndrome, that once caused permanent blindness, deafness and other physical or mental impairments, have virtually been conquered in the United States and other industrialised nations; yet there persists a current epidemic of a condition referred to as Autism Spectrum Disorder. Even though the call for children’s rights has not always been heard, and movements have faltered, a pattern of progress is suggested and expressed in social policy and in new paradigms for understanding exceptionality and childhood. Disability is, and likely to remain, a historically situated social construction.

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