

# CHAPTER 118

## GENERAL DISABILITY: THE CONCEPTS

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### Introduction

*Webster's New World Dictionary of American English*<sup>1</sup> makes the following distinctions of terms related to general disability:

**Disable:** (1) to make unable, unfit, or ineffective; cripple; incapacitate; (2) to make legally incapable; disqualify legally

**Disability:** (1) a disabled condition; (2) that which disabled, as an illness, injury, or physical handicap; (3) a legal disqualification or incapacity; (4) something that restricts; limitation; disadvantage

**Cripple:** (1) a person or animal that is lame or otherwise disabled in a way that prevents normal motion of the limbs or body; somewhat offensive when used to refer to a person

**Handicap:** (2a) something that hampers a person; disadvantage; hindrance; (2b) physical disability

In the present age of “enlightenment”, many terms have been suggested to describe a person with a disability. However, the terminology used can be politically sensitive and abruptly change from decade to decade. For example, should we say “disabled child” or “child with a disability”? While remaining sensitive to others, we must also utilise terms that can be understood by the audience to whom we address these terms. When trying to describe the work being done in Africa, compromise may be necessary. When directing letters to friends in North America and referring to working with the handicapped, I often receive inquiries about whether these were people with alcohol- or drug-related problems. One hospital in Africa providing care for children with disabilities was named Bethany Crippled Children’s Centre. Many were aghast that the word “crippled” had been used with such indiscretion. The explanation usually given was that the name was an attempt to communicate the type of work being done to the “least educated in the most rural village”. Disabled, handicapped, and various other terms did not communicate to these nearly as effectively as “crippled”, which seemingly was a term that many understood.

### Demographics

Some studies estimate that 1% of the people in the world are disabled.<sup>2</sup> Africa is likely a different scene, however. Due to wars, civil strife, various religious beliefs, economic constraints, a relative lack of medical care, and a greater incidence of infectious diseases, the incidence of disabilities in Africa is likely greater than 1%. Some estimate an incidence of as low as 3%, whereas others, such as Dr. Rodney L. Belcher, the former chief of orthopaedics at Mengo Hospital in Kampala, Uganda, have suggested that the figure is actually as high as 10%. Regardless of the exact number, it is likely that the incidence of disabilities in Africa is greater than for the world in general. In the early 1990s, Dr. Belcher estimated that 10% of the population of Uganda had some disability; he also commented: “Each week in Kampala we have approximately three new cases of culture-proven polio.” The level of disability in that country was undoubtedly related to nearly two decades of civil strife and the breakdown of the economic and health systems.

The Kenya scene in 2008 is far different in certain respects from that of many other countries. Even though the number of medical practitioners is large by sub-Saharan African standards, it falls far short of that of nearly all Western countries. Polio is rarely seen in its acute phase, even though it probably is not eradicated. The incidence of talipes, spina bifida, cleft lips and palates, and various other congenital abnormalities in Kenya is not known, but it would appear that their presence is as plentiful as it is in other African countries. There are more than 40 tribal groups in Kenya, and patients with clubfeet, spina bifida, and cleft lips come from nearly all of those tribal groups.

Children with disabilities seem to abound in nearly all sub-Saharan African countries. Few have been adequately treated, and most have had no treatment.

### Aetiology

Disabilities in general can be identified as either congenital or acquired. One could further break down the congenital disabilities into those of genetic aetiologies (e.g., Down syndrome); those related to a nutritional deficit (e.g., neural tube defects); those associated with some inciting event (e.g., rubella); and random expression. Moreover, some defects that may appear identical could be, in reality, either “familial” or ran-

Table 118.1: Recurrence risks for some defects.

Defect	Normal parent of one affected child*	Recurrence risk for:	
		Future males	Future females
Cleft lip with or without cleft palate	4–5%		
Cleft palate alone	2–6%		
Cardiac defect (common type)	3–4%		
Pyloric stenosis	3%	4%	2.4%
Hirschsprung's anomaly	3–5%		
Clubfoot	2–8%		
Dislocation of hip	3–4%	0.5%	6.3%
Neural tube defects—anecephaly, meningomyelocele	3–5%		
Scoliosis	10–15%		

Source: Jones KL. *Smith's Recognizable Patterns of Human Malformation*. Saunders, 1997, Table 4-1.

\*Range of recurrence risks observed.

Table 118.2: Ratio of males versus females for various disorders.

Disorder	Male-to-female ratio
Pyloric stenosis	5:1
Clubfoot	2:1
Cleft lip with and without palate	2:1
Cleft palate alone	1:1.3
Meningomyelocele	1:1.5
Anencephaly	1:3
Congenital dislocated hip	1:5.5

Source: Jones KL. Smith's Recognizable Patterns of Human Malformation. Saunders, 1997, Figure 4-14.

dom. Among family members it is not uncommon for more than one member to be affected with clubfeet, cleft lips, cardiac defects, pyloric stenosis, Hirschsprung's disease, hip dislocation, neural tube defects, scoliosis, or other problems (Table 118.1).<sup>3</sup> The gender distribution may also vary in various malformations (Table 118.2).<sup>3</sup>

One of the most commonly identified defects related to a nutritional deficit are neural tube defects. A deficit of folate creates an environment in which this defect is manifested more commonly. In countries where nutritional fortification with folate has occurred, the incidence of this problem has diminished by 66–75%. In North America, the incidence of children born with spina bifida has diminished due to folate fortification. The incidence of children conceived who have spina bifida may be considerably underreported, however, due to the frequent abortion of fetuses identified by ultrasonography (US) as having neural tube defects.

Other disabilities, such as burn contractures and polio, are generally preventable either by improved living standards or immunisation. Improved community education should promote the reduction of many disabilities as people learn basic steps they can take to prevent disabilities from occurring. Genetic counselling can also assist in diminishing the number of children born with disabilities.

## Clinical Presentation

### History

A good history and physical examination usually will lead to an accurate diagnosis and can often save time and money. A disproportionate number of those described as disabled come from families who are poor and/or less educated. Many, if not most, of these children are born at home with, at best, a traditional birth attendant (TBA) in attendance. Most newborns will not have had that early evaluation, even briefly, that usually occurs in a hospital birth; this initial physical can lead to early diagnosis and a better informed attempt at seeking appropriate care. Often, these families have little access to appropriate medical care, even if they were to recognise that “something can be done”.

Parents of children from poor and less educated families will also frequently not understand the importance of immunisations and early care.

### Physical Examination

The variation in physical findings parallels the diseases encountered and the timing at which the child arrives for care. Burns, spina bifida, hydrocephalus, clubfeet, polio, and the various other disabilities often arrive late in the course of their disease.

A routine physical examination will easily identify most of the overt disabilities, but the less obvious associated conditions (e.g., cardiac, renal, and liver problems) often go undiscovered longer, frequently complicating their management.

## Investigations

In the case of late arrival, the diagnosis often declares itself. Yet, for certain infants, a differentiation of aetiology must be determined. An example of this is the clinical presentation of a child with clubfeet. In differentiating among the congenital clubfoot, a nearly identical foot with polio, and the “clubfoot” associated with spina bifida, a few

questions can clarify the aetiology: “Was the child born with this deformity?” “At what age did the child demonstrate this deformity?” “Does the child seem to have sensation and motor activity in the leg?”

In other situations—for example, in the hydrocephalic patient—one might seek to identify the aetiology of the condition (i.e., infectious, congenital, or other). The aetiology may dictate the extent of brain damage and what can, or should, be done. If the fontanelle is open, ultrasound (US) can clarify the size of the ventricles as well as identify the amount of cortex; this may suggest the potential long-term outcome. US is relatively inexpensive, whereas computed tomography (CT) and magnetic resonance imaging (MRI) can be costly. Often, however, none of these exams are available. If they are unavailable, in the case of hydrocephalus, the head circumference should be measured and compared to the normal range for that age (Figure 118.1). The size of the fontanelle, together with the history, and general impression are important. In most cases, this is sufficient to determine the presence of hydrocephalus.

In the case of an occipital encephalocele, US may help clarify the contents and the potential viability of the infant if this is to be repaired; the size of the ventricles is also important to determine whether there is an accompanying hydrocephalus needing shunting.

A good examination of patients with congenital talipes equinovarus (CTEV) can suggest the best pathway for treatment without expensive tests. Similarly, a close examination of the deformities and motor activity of the child with polio should suggest the way to proceed. In all of these cases, the parent and/or patient needs to understand what can and cannot be done, as well as the ultimate expectations.

## Management

Many African countries have few specialists capable of meeting the needs of the disabled children. If the specialists are available, they are almost never present in sufficient numbers. Poor families often lack sufficient funds to access the care, even if it is available. Many who do seek care come late—often too late.

The setting in which the patient presents, the level of expertise of the surgeon, the availability of materials, the investigative possibilities, the financial status of the patients, the ability to provide follow-up care, and various other factors often will dictate the potential management and outcome. The outcome in a limited-resource setting, although possibly far inferior to that anticipated in a sophisticated medical setting, might be considered quite acceptable to many in Africa. The expectations may be quite variable. Function, cosmesis, and/or practicality are all important; however, the expectations of the parents and/or the child should be ascertained prior to surgical care. A careful explanation of the procedure, the postoperative care, the need for appliances postoperatively, and the anticipated functional ability will assist in avoiding misunderstandings.

In the case of talipes, management should begin shortly after birth with manipulation and plaster. Early evaluation of the foot will allow the surgeon to explain the plan and the expectations to the parents. The Ponseti technique of serial casting and appropriate intervention will often avoid major surgical procedures. However, this technique demands that the parent bring the child early and cooperate in the care of the child. Such consistent care is often not possible for these children because the family does not understand the importance of the consistency of care, or they may live in a remote location without access to the health care workers needed to provide this care, or they cannot afford this level of care. Also, in reality, the regimen suggested by Ponseti is unknown to most of the health care workers of Africa, and, if known, cannot be provided due to a lack of training and/or supplies. In a significant number of the children treated by this method major surgery will still be necessary.

Often in East Africa, children with club feet arrive for care when they are several years of age or even older; in most of these cases, there has been no previous orthopaedic care. First, the surgeon must decide

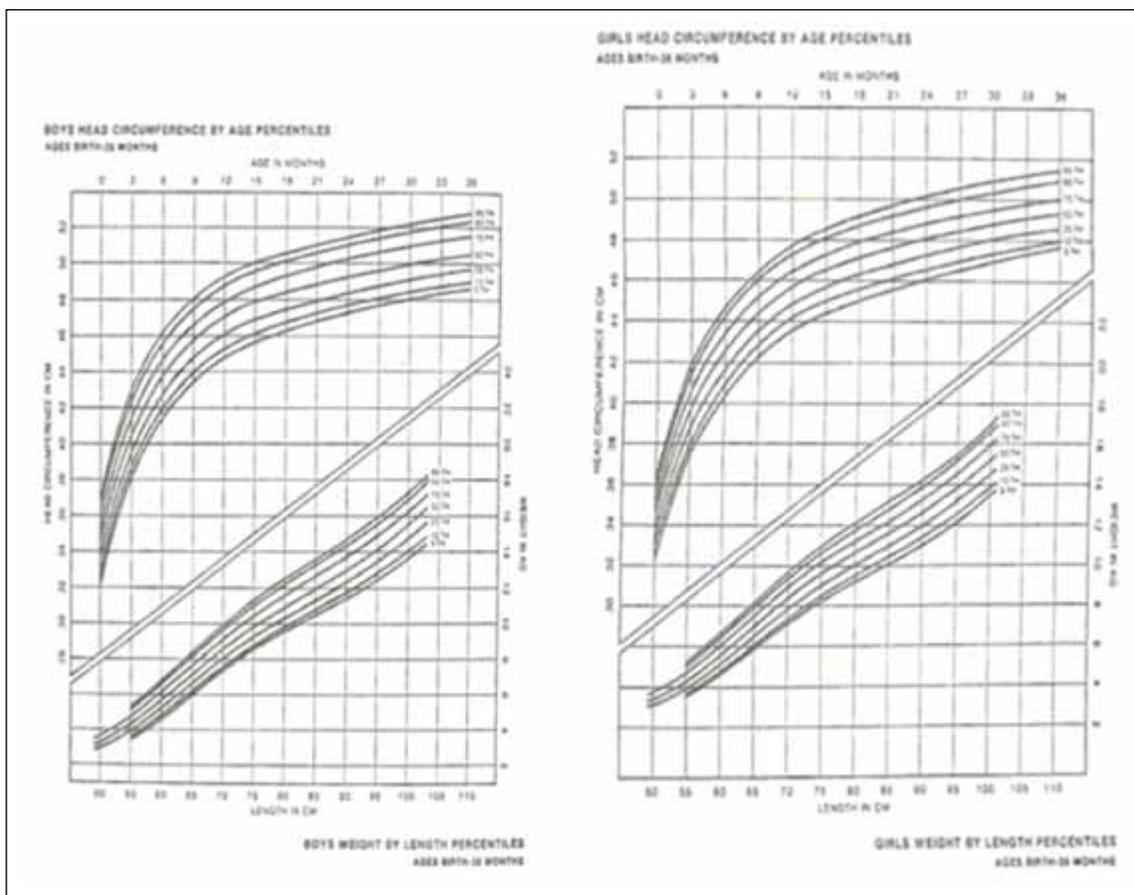


Figure 118.1: Growth charts for boys (left) and girls (right) from birth to 36 months: head circumference by age and weight by length.

Source: Modified from Hamill PV, et al. Physical growth: National Center for Health Statistics percentiles. *Am J Clin Nutr* 1979; 32(3):607-629.

whether any operation should be performed at that time, and, in the case of the immature foot, whether a soft tissue release should be done while delaying operations on the bony structures until the foot is more mature. One must seriously question whether the outcome will be a more functional foot, whether the child will wear a caliper (brace), and/or whether the child will return for further care at an older age.

In cases of congenital pseudarthrosis of the tibia, once again, one must consider the setting. In the West, an amputation with prosthesis may be the preferred pathway; in an African setting, however, attempts at repairing the pseudarthrosis, even if it results in a short leg, may be preferred due to the lack of available prostheses, especially for a growing child, as well as the expense of providing these prostheses.

The late arrival for treatment of many children may dictate operative and nonoperative solutions that frequently are not described in most Western textbooks. Usually, Western medical texts assume that early care can be or has been provided. In the case of late presentation, the results of any attempt at treatment are often fraught with more complications and usually inferior to those to be expected if early care had been sought.

Some situations, possibly particularly in the case of polio deformities, may dictate consideration of remedial operations with, or without, bracing. Assuming inadequate follow-up and possibly poor compliance, a remedial operation may be the best option. A “perfect” operation without the use of a brace will often fail. Also, a growing child with a caliper will need to have the caliper replaced.

Obviously, the usual approaches and expectations must be modified when confronted by a 20-year-old with a cleft lip and palate, a 9-year-old with an untreated clubfoot, an 8-year-old with severe rickets, a 5-year-old with advanced hydrocephalus, or burn contractures untreated for decades. The contextualized treatment must consider not

only the surgical skills and treatment environment but also the abilities of the nonphysician staff to maximise the potential created by the surgical team. The ability to apply such modifications may allow the child to enjoy a much improved quality of life.

In some remote settings the health care workers (HCWs) may be the *last option* for many children. These HCWs often will be satisfied with less than “state-of-the-art” techniques, and many are just looking for a better “quality of life”. Although such HCWs are not specialists, some who have some surgical experience may be put in a situations where the child has no other options. Many good books that are often considered remedial or out of date by the high-level specialist may offer good pictures and a description of the procedure that can be followed. These books (e.g., Lehman’s *The Clubfoot*;<sup>5</sup> Goldstein’s *Atlas of Orthopaedic Surgery*;<sup>6</sup> and Tachdjian’s *Pediatric Orthopedics*;<sup>7</sup>), often open and sitting by an operating table, lend support to the desperate surgeon. Many techniques of previous decades offer solutions that are quite satisfactory. Some books (e.g., Staheli’s *Fundamentals of Pediatric Orthopedics*,<sup>8</sup> *Practice of Pediatric Orthopedics*,<sup>9</sup> and *Arthrogyposis: A Text Atlas*<sup>10</sup>) make difficult explanations simple enough for the generalist. Much of what is included in these books can also be obtained online.

Albright’s *Operative Techniques in Pediatric Neurosurgery*<sup>11</sup> can be instructive for many surgeons. If one can shunt a child with hydrocephalus or close the back of a child with spina bifida, a large number of children can be assisted. Many varieties of ventriculoperitoneal shunt are available, with great variations in cost; Warf has found the Chhabra™ ventriculoperitoneal shunt to be equally successful to expensive Western shunts.<sup>12</sup> (In addition, the Chhabra shunt has been quite useful as an indwelling catheter in hypospadias repairs. It is soft silastic, inert, and of just the right size for the average older child.)

## Postoperative Complications

When children with disabilities arrive late in the course of their disease, it is often more difficult to attain the desired endpoint. Too much done too quickly will often encourage complications. In the developing world, staged procedures may lead to fewer complications. The staged approach is often wisest with imperforate anus, hypospadias, neglected clubfeet, some polio deformities, osteogenesis imperfecta, spina bifida associated with hydrocephalus, and various other disabilities. For some disabilities, such as open spina bifida, infections will occur more commonly when the arrival of the child is delayed. In some cases, a further delay in surgical care to allow for improved preoperative preparation, intravenous antibiotics, and so forth may be best. Part of this preparation may include thorough cleansing of a dirty wound, the correction of anaemia, and malnutrition, and the preparation of the parent.

Also, having too few trained medical staff can lead to complications. When operative care includes plaster, the staff may either not be able to evaluate the extremities as often as desired, or not have adequate understanding of the importance of this task. Sometimes, it may be best to compromise by utilising a splint instead of a cylinder plaster.

Often, outcomes are compromised by a lack of specialised personnel. It is essential that an active therapy plan and program be in place following specialised operations on the hands of most burn victims. A planned, and sometimes painful, exercise program can make the difference between failure and success. Specialised personnel that are often helpful include orthopaedic technologists, speech therapists, occupational therapists, physiotherapists, and dentists.

## Prognosis and Outcomes

Both prognosis and outcome are complicated by late presentation and preoperative conditions that contribute to poor outcomes. In addition to late presentation are the conditions dictated by the setting and expertise of those caring for the patients. Many of the factors expected in the West (e.g., surgical specialists and subspecialists in a wide variety of fields, greater expertise in nursing, good physiotherapy and occupational therapy, state-of-the-art orthotics and prostheses, sophisticated operative settings, and many other factors) are often not present in the developing world; perhaps contributing to less desirable outcomes. One might add that refugee status and a transient patient population also may contribute to poor outcomes. Often these patients do not understand the preoperative, operative and postoperative plans; some have no choice in their ability to regularly access appropriate medical care. The language barrier in many settings may promote unrealistic expectations on the part of the patient and parents and later disappointment.

In the midst of all of the difficulties, it is important for health care workers to become familiar with the pathways that may be acceptable to their patients and their families. A relatively crude therapy device that may be difficult to accept in the West is often quite acceptable in African settings. Werner's *Disabled Village Children*,<sup>13</sup> an important guide to the diagnosis and treatment pathways of various disabilities, also has designs for appliances that are quite effective.

In the case of cancer and the need for radiotherapy and/or chemotherapy, however, the cost is often prohibitive and specialised care is unavailable.

## Prevention

Even though Western Hemisphere data suggests that polio has been eliminated there, this is not true in Africa. War, civil strife, and, in some cases, religious bias have led to fewer preventive measures in economies that may be oriented towards other goals. Within eastern and northeastern Africa, polio is more prevalent in Sudan, Ethiopia, and Somalia. Two decades ago, Uganda could have been included in this list due to its own civil strife. Immunisation, vitamin A, adequate teaching about various diseases, preconception folate, genetic counselling, antihelminthics, and appropriate iodine, as examples, would help in the prevention of various disabilities. One could easily add to this list clean, adequate water and soap as well as effective toilet facilities as contributing factors in a "preventive" program.

## Ethical Issues

Children with disabilities often enter school later than other children. Unfortunately, many special schools are insufficiently staffed and are not equipped for the numbers and types of children coming for training. Most of these children need not only the normal education of children without disabilities, but also therapy and basic education in living skills. Few, if any, African countries have sufficient funding and expertise to provide such an educational experience for these children.

Some disabled children continue to be considered a "curse" on the African continent. They may be a shame to their family, their village, and/or their country. Consequently, many of these children are hidden from public view, sometimes for years. This attitude, a part of the cultures of many people, is more common in remote village settings where there is little education about medical issues and other problems. Such an attitude delays or prevents treatment. Often, when a choice—economic, cultural, or other—has to be made by the family, the child with a disability does not receive care. Frequently, this is due to economic issues within a poor family. Obviously, some disabilities are more overt and others are unseen. In general, the view that disabilities are a sign of a curse is still widespread in Africa.

In sub-Saharan Africa, where the economies of most countries are seriously compromised and there is an abundance of very poor people, the questions of what "should be done" often emerge. What are the circumstances that "allow" an investment in a child with hydrocephalus or spina bifida with all of the considerations that go along with their long-term care? What can the economy "afford" in its investment in disabled children in general? As physicians, what is our responsibility in pursuing the care of these children? These are issues for consideration; the answers likely will vary from country to country and from family to family.

### Key Summary Points

1. Children with disabilities in Africa tend to arrive later in the course of their disease and will often need care and procedures not necessarily described in Western textbooks.
2. The parents of children should be carefully advised about the prognosis and expectations for their children.
3. When beginning work with children with hydrocephalus and spina bifida, the physician should select carefully the cases on which to operate. The physician should incorporate a team that includes an operating theatre technician, a physician with surgical skills, and a special nurse for pre- and postoperative care. A careful look at all of the ramifications of the care of these children is warranted.
4. Specialised surgeons are not available in most of Africa. Physicians must seek advice and do their best.
5. Regular follow-up is recommended for nearly all disabled children. Their needs will likely change as they get older.
6. The physician must recognise the setting and the audience and tailor the investigation and care to the patient's abilities. Although the hospital may not be "state-of-the-art", the patients also are likely to be poor.
7. Even older surgical textbooks are of value. They may include simpler operations that will help most of the children and have techniques that are much more inviting than those in sophisticated current textbooks.
8. The health care worker may be the only hope for many of the parents and their children.
9. Many specialists in the world might be willing to come to the local hospital and teach. Invite them.

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