

The right of persons with disabilities to the highest attainable standard of health

IF Response to the consultation launched by
the UN Special Rapporteur on the Rights of Persons with Disabilities

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Introduction

The [International Federation for Spina Bifida and Hydrocephalus \(IF\)](#) is a global organisation of persons with disabilities governed by adults with spina bifida and/or hydrocephalus (SBH), or parents of children with SBH. IF's mission is to improve the quality of life of people with spina bifida and hydrocephalus and their families, and to reduce the incidence of neural tube defects and hydrocephalus by primary prevention; by raising awareness and through political advocacy, research, community building, and human rights education. Universal respect of the rights reaffirmed in the UN Convention on the Rights of Persons with Disabilities (UNCRPD) for all children and adults with SBH is IF's underlying philosophical base, and we support the call to leave no one behind through the implementation of the Sustainable Development Goals (SDGs).

IF is a member of International Disability Alliance, the European Disability Forum and the International Disability and Development Consortium, as well as EURORDIS and Rare Diseases International. IF has held consultative status to ECOSOC since 1991.

Spina bifida is one of the most complex neural tube birth defects compatible with life, characterised by various degrees of damage to the spinal cord and consequent life-long health conditions necessitating care and support related to reduced mobility, urological and bowel management issues, orthotic needs, and weight management. Many people with spina bifida also develop **hydrocephalus**, which is an accumulation of excess cerebrospinal fluid in the brain. If untreated, hydrocephalus can cause blindness, intellectual disabilities, and premature death. Although those affected are usually born with hydrocephalus, a person can also develop the condition from neonatal infection, tumors, hemorrhage, etc.

Although persons with SBH may have similar healthcare needs as persons without disabilities, they also experience distinct needs that stem specifically from the complex nature of the disabilities in question. Their care requires an **integrated, multidisciplinary approach, throughout the lifespan**, which focuses not only on their physical health, but also their mental and social well-being. In particular for young people with SBH, the proper preparation for a smooth **transition from paediatric to adult care** is of utmost importance.

An in depth description of the needs of persons with SBH throughout the life cycle can be found in our response to the consultation launched by the UN Special Rapporteur on the Rights of Persons with Disabilities in 2016, on the provision of support to persons with disabilities.¹

Through feedback from our members, and repeated requests for help from individuals, we know that many people with SBH are lacking access to this type of holistic care and support, as well as access to the necessary medical and assistive devices and products, and medication, in various parts of the world.

Recalling the 2011 World Report on Disability, we would also like to emphasise here that a wide range of factors determine health status, including individual factors, living and working conditions, general socioeconomic, cultural and environmental conditions, as well as access to healthcare services.²

Also, the World Health Assembly resolution 63.17 on Birth Defects³ (since then, WHO uses the term “congenital anomalies”) called upon its members to include children born with disabilities in the health care systems, to support families and raise awareness. This specific resolution also recalled WHA resolution 58.31, calling for universal health coverage, which we also support as mechanism to include the healthcare needs for all.

Underlying obstacles to achieving the highest attainable standard of health

Lack of data

Data with regard to the prevalence of SBH is incomplete or lacking. In developing countries, children with these disabilities are often not registered at birth and parents may not seek healthcare, but instead hide them from view due to **stigma and superstition**. In countries that do have registries for the epidemiologic surveillance of congenital anomalies such as SBH, **data may only represent limited parts of a country** and exclude densely populated urban areas. When newborns with disabilities do receive care, it is difficult to find statistical data on patients with SBH, the treatments that they receive, and their outcomes, as specific SBH patient registries barely exist.

The lack of data with regard to newborns with SBH and people living with the conditions can lead to the wrongful presumptions that SBH are **rare conditions**, and as such, not worthy of investment in medical research or development of adequate healthcare services.

As a result, research into the appropriateness and effectiveness of the treatment and care that people with SBH are receiving worldwide is mostly unavailable.

¹ [IF submission on support to persons with disabilities](#) (2016)

² [WHO World Report on Disability](#) (2011)

³ http://apps.who.int/gb/ebwha/pdf_files/WHA63/A63_R17-en.pdf (2010)

We know, from studies, that the incidence in pregnancy ranges between 1/1000 in developed countries to 20/1000 in regions with specific nutrition problems.⁴ On average we can assume 3/1000. Most of these children will have died before the age of 5 because of lack of care⁵.

In line with the UNCRPD and SDG 16, target 16.9, **registration at birth** of all children, including children with congenital anomalies and disabilities, is required. Furthermore, **SBH patient registries** need to be set up to gather data on long-term outcomes of the available treatment and care. Having access to correct data will help countries to plan for care, improve healthcare and support services, and to avoid preventable additional health issues for people with SBH.

In the United States, a [National Spina Bifida Patient Registry](#) was set up in 2008 to collect the scientific data needed to evaluate existing medical services for spina bifida patients, after advocacy efforts of our member the Spina Bifida Association (SBA). The Registry provides the framework for a systematic approach to improving the quality of care received at spina bifida clinics nationwide. It collects information from patients that visit specific SB clinics in the US to understand the associations between medical procedures and health outcomes. Clinics must meet specific requirements, which include the number of new spina bifida patients they see each year, and their staffing capacity.

The first steps have been taken by the government in Italy to set up a [National Spina Bifida Registry](#), by signing agreements with our member ASBI and local SB associations. Next, a contract will be signed with the Higher Health Institute to make this registry a reality. These two projects are excellent examples of close cooperation between government organisations and SB associations. Patient involvement in policy measures that have an impact on their quality of care and their quality of life is key.

Lack of knowledge and persisting stigma and misconceptions

Healthcare providers may only see few patients with SBH on an annual basis throughout their career. This is either because people cannot or do not seek or reach healthcare services, or because the prevalence is low as a result of primary prevention measures, or due to **prenatal diagnosis and termination of pregnancy**. As a result, they will have limited knowledge and experience based on single cases.

In addition, medical literature used in healthcare education can be **out of date and biased**, and may not include the latest information on the correct treatment of SBH and the improved expected outcomes that can be achieved with timely, appropriate, multidisciplinary care throughout the various stages of life.

⁴ [Describing the Prevalence of Neural Tube Defects Worldwide: A Systematic Literature Review](#)

⁵ [Estimates of global and regional prevalence of neural tube defects for 2015: a systematic analysis](#)

Consequently, inaccurate and often dangerous speculations continue to be made about the **future quality of life** of those born with SBH, often leading to denial of essential treatment and care that would help people with SBH to survive and thrive. Cultural superstition with regard to children born with disabilities can have a similar devastating impact.

These negative assumptions with regard to SBH can also cause **stigma and discrimination by association**⁶, affecting parents(-to-be). Parents that choose to keep their child may have difficulty getting access to healthcare for their baby, may be criticised by society for creating a “burden” on the healthcare system and social services, or may be shunned and excluded from their community for having a child with a disability.

Where children do receive the appropriate care, they can live into adulthood. The current generation of adults living with SBH often consider themselves as “pioneers”, as there is a lack of medical research into **ageing with SBH**, for instance the long-term effects of childhood procedures, or lack thereof.

There is also very little information about **SBH and parenthood**. This may in part be due to the wrongful assumption that people with SBH cannot conceive. As a result, it is typical for sexual and reproductive health services to be unable to provide people with SBH with correct information and the necessary support.

The difficulties of searching for and getting access to appropriate information and services that are specific to their needs can have a negative impact on the mental health of people with SBH.

Up-to-date information and international guidelines are needed for the treatment and care of SBH from before birth to adulthood, these should be accessible to healthcare providers, to people with SBH, and to parents(-to-be) worldwide. Additionally, in order to remove attitudinal barriers, countries need to raise awareness on the rights of persons with disabilities, throughout the whole of society.

Guidelines, information, and awareness raising policies need to be developed in close cooperation with SBH associations and other organisations of persons with disabilities, in line with the UNCRPD.

An example of information provision to parents and people with SBH is the dedicated website which was launched by our member Spina Bifida Hydrocephalus Scotland, “[Hydrocephalus and young people](#)”. It covers topics such as causes, treatment, living with the condition, impact on cognition and executive function, and how to address various challenges, in clear and understandable language. The website was made possible through grants of the Big Lottery Fund and RS MacDonald, yet initiatives such as this should not have to rely on charity, but should be supported by national governments.

⁶ an illustration of stigma and the potential consequences on access to health care can be found in this documentary shot at one of the IF partner hospitals: <https://vimeo.com/48893434>

In Argentina, our member APEBI was able to create a brochure in plain language with extensive information on Spina Bifida throughout a person's life with the support of the Programme for Rare Diseases of the Ministry of Health, "[Aportes sobre espina bífida - A lo largo de la vida](#)".

Another of our members, the Spina Bifida Association in the United States, has been developing [Spina Bifida Healthcare Guidelines](#), in close cooperation with the US government's Centers for Disease Control and Prevention (CDC). The guidelines will be published in May this year. Experts from various hospitals and countries have been involved in the writing of these guidelines and their extensive review. The project has been supported by the Spina Bifida Collaborative Care Network.

Lack of accessibility

Access to healthcare is a broad concept. It can for instance be determined by availability, affordability, appropriateness, geographical location, specific barriers for people with disabilities (lack of wheelchair ramps, discriminatory attitudes of healthcare providers due to stigma and prejudice, lack of information in accessible formats, inaccessible buildings or medical equipment, etc.), and the ability of patients to seek healthcare.

As explained above, the required access to integrated multidisciplinary care for people with SBH throughout the life span simply does not exist in many countries, due to lack of data, lack of knowledge, and/or lack of investment. In low and middle income countries the lack of financial resources may be an additional barrier for the development of and investment in appropriate and accessible healthcare services for persons with disabilities. Yet even with limited means, good care is possible.

For its projects in African countries, IF developed the **Spina Bifida and Hydrocephalus Interdisciplinary Program (SHIP)**⁷, which offers a coordinated, multidisciplinary approach towards an individualized care program. The objective of SHIP is to involve all stakeholders, such as (health)care providers and parents, and to improve collaboration in order to ensure continuity and quality of care. To this end, SHIP Coordinators are appointed at each SHIP location, who collect and disseminate data, knowledge, and information to all parties involved. Each child is provided with a SHIP passport with an overview of the most important data, e.g. emergency contact details, medical history, medication use, etc., which stays with the child and needs to be presented and updated at each hospital visit. SHIP Protocols and leaflets on various topics related to SBH have been developed.

In the European context, IF research conducted in 2016 among its EU membership showed that limited access to multidisciplinary care for SBH is closely connected with **long waiting**

⁷ [SHIP Presentation slides](#)

times for individual specialist consultations. Of the respondents, 57% had to wait 1-12 months before receiving an appointment, and 5% had to wait for over a year⁸. Apart from the risks of (secondary) health conditions not being diagnosed and treated in time, regularly **needing time off for multiple appointments with different healthcare providers** can severely disrupt the education of young people with SBH and have a negative impact on employment of (young) adults with SBH and of parents of children with SBH.

In countries that do offer multidisciplinary care for SBH, or when cross-border healthcare is an option, the services may still be inaccessible to many, due to the **distance to hospital or clinic**, the lack of **affordable and accessible transport**, and the additional **costs of accommodation**. These costs can be significant for families, or for persons with SBH who travel with a personal assistant, especially when we remember that they are not properly covered by healthcare insurance or other social protection measures.

Another result of the 2016 IF research is that 41% of respondents mentioned that their or their child's **medical expenses were only partially covered by the state**, while 6% stated that they do not have any coverage at all. These costs of care do not only apply to treatment, but also to medical devices, e.g. shunts for the treatment of hydrocephalus, medical products such as urinary catheters, and assistive products⁹ (crutches, braces, orthoses, wheelchairs, absorbent incontinence products, etc.), and medication (for instance antibiotics, Oxybutynin, Botox).

Lack of funds can result in delays in treatment so that the outcomes are worse than if the treatment was possible much sooner. In Uganda, funds for the issues as simple as bus fare might result in a delay of hydrocephalus treatment for six months. Often artificial monetary barriers are created by asking unnecessary procedures to take place beforehand, such as CT-scans.

Based on anecdotal evidence, for instance through individual requests for help and various news articles, IF has learned that parents can often not afford treatment when their child has hydrocephalus, due to the cost of the shunt, the necessary surgery, and the hospital stay. In cooperation with its partners, **IF donates Chhabra shunts** to its projects in developing countries. A 2005 study showed that this affordable shunt system works just as well as the far more expensive shunts that are used more often.¹⁰ Some SBH parent groups in our projects also developed a so called "House of Hope", close to the hospital, where children with SBH and their parents can stay after surgery. This reduces the need for additional travelling, it lowers the costs of accommodation, and it allows for quick access to the hospital in case of an emergency.

⁸ [Right to health: Reality of persons with Spina Bifida and Hydrocephalus \(2016\)](#)

⁹ [WHO Priority Assistive Products List \(APL\)](#)

¹⁰ [Comparison of 1-year outcomes for the Chhabra and Codman-Hakim Micro Precision shunt systems in Uganda: a prospective study in 195 children](#)

The lack of access to multidisciplinary SBH care and lack of universal health coverage can create a lasting financial burden for persons with SBH and for families with children with SBH, as the need for follow up care and support, and access to medical products, medication, and assistive products, is lifelong.

Novel procedures

Nowadays, parents-to-be who want to achieve the highest attainable standard of health for the child with SB that they are expecting, may want to have access to **foetal surgery for SB**. According to scientific evidence, this high risk procedure, for both mother and foetus, offers a significantly reduced risk of hydrocephalus, and a chance at improved mobility, compared to newborns with SB who are treated after birth. Parents-to-be are often highly interested in any treatment that may offer their child the best start in life, even if there are risks and even if there is the possibility that their child may still need a shunt, or will not belong to the group with improved mobility.

As this type of surgery requires high levels of expertise and a great amount of hospital resources, both with regard to prenatal screening and diagnosis and the operation itself, it is only available in a limited number of countries. Also, only a small group of patients will qualify, due to the strict selection criteria for both mother-to-be and foetus, and the limited timeframe in which the procedure is possible. Concentrating this treatment option in a few selected hospitals will ensure that healthcare professionals will perform enough foetal surgeries for SB to maintain and improve their skills.

However, this limited availability creates the additional costs of travel and accommodation, mentioned in “*Lack of accessibility*”, which can create a barrier. Additionally, in some countries the procedure itself can still be considered “experimental” and will not be covered by healthcare insurance. This can create a huge financial burden for parents-to-be, who may take out a loan or mortgage their home. Some may turn to online fundraising campaigns, but after diagnosis and qualifying for foetal surgery for SB, there are only a few weeks left before the procedure needs to be done. When they can’t raise the money for novel treatments, parents will have the additional burden of feeling they haven’t done all they can for their child. Foetal surgery for SB also requires a **multidisciplinary approach**. Among others, it includes paediatric surgeons, foetal medicine specialists, anaesthetists, obstetricians, and nurses, and it requires adequate counselling of, and support for, parents-to-be.

A similar lack of availability exists for the treatment of infant hydrocephalus through a procedure called combined Endoscopic Third Ventriculostomy and Choroid Plexus Cauterization (ETV/CPC). While this procedure is technically more difficult than shunt placement and requires extensive training and access to a neuro-endoscope, it takes away the need for a shunt and removes the risk of shunt failure and the need for shunt revisions. At the same time, the cognitive outcomes of children treated with ETV/CPC are similar to children that are treated with a shunt.¹¹

¹¹ [One-time hydrocephalus operation, alternative to shunting, brings good outcomes for babies](#)

Access to regular healthcare

From responses on surveys and anecdotal evidence, it is clear that persons with SBH do not receive the same general healthcare as their peers. In developing countries, we see gross malnutrition, denial of malaria treatment, reduced vaccination etc. because of discrimination and stigma for children born with disabilities. Persons with SBH report postponing family doctor visits because of the serious financial burden that the specialty services already place on families. Although not the subject of this paper, which focuses on disability-specific healthcare, we call upon the Special Rapporteur to also look at discrimination within regular healthcare systems.

Summary

To reach the highest attainable standard of health for people with SBH, it is key that their existence is acknowledged, through registration at birth and by setting up SBH patient registries. Next, they should have access to integrated, multidisciplinary care, throughout the lifespan, which focuses on all aspects of health, including sexual and reproductive health, and ageing with SBH. Their medical needs must be met, but their mental health and social well-being must be taken into account as well. Apart from specialist care, regular healthcare, such as family doctors, needs to be accessible too.

Data from patient registries needs to be monitored and evaluated to ensure that patients with SBH receive the best possible treatment and care. Best practices need to be developed into protocols and guidelines, so that unnecessary procedures can be avoided. Up-to-date information needs to be easily accessible to everyone.

To protect people with SBH against misconceptions, prejudice, stigma and superstition, which create barriers to accessing healthcare services and have an overall negative impact on their quality of life, countries need to raise awareness on the rights of persons with disabilities, throughout the whole of society.

Universal health coverage and social protection measures need to be in place, to ensure that every person with SBH can have access to the healthcare and rehabilitation services that they are entitled to, including the regular healthcare available to their peers. Arising from our experience in low-resource situations, we know that funding is often the issue given to justify the discrimination and health inequalities we see now. We fundamentally disagree with this reality and believe that the rights of those living with disabilities should be pursued regardless of these socio-economic factors. We do acknowledge that it is important not to fall in the trap of expensive care, as that risks to become an extra barrier for access to care. Many low-cost treatments with significant impact on the quality of life have a proven track record, as we have shown and disseminated.



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We wholeheartedly thank you for taking on this enormous task of reporting the situation of access to care for persons with disabilities and for the opportunity of reporting our findings to you.

For the board of directors and the global expert panel of the International Federation for Spina Bifida and Hydrocephalus,

Warm regards,

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