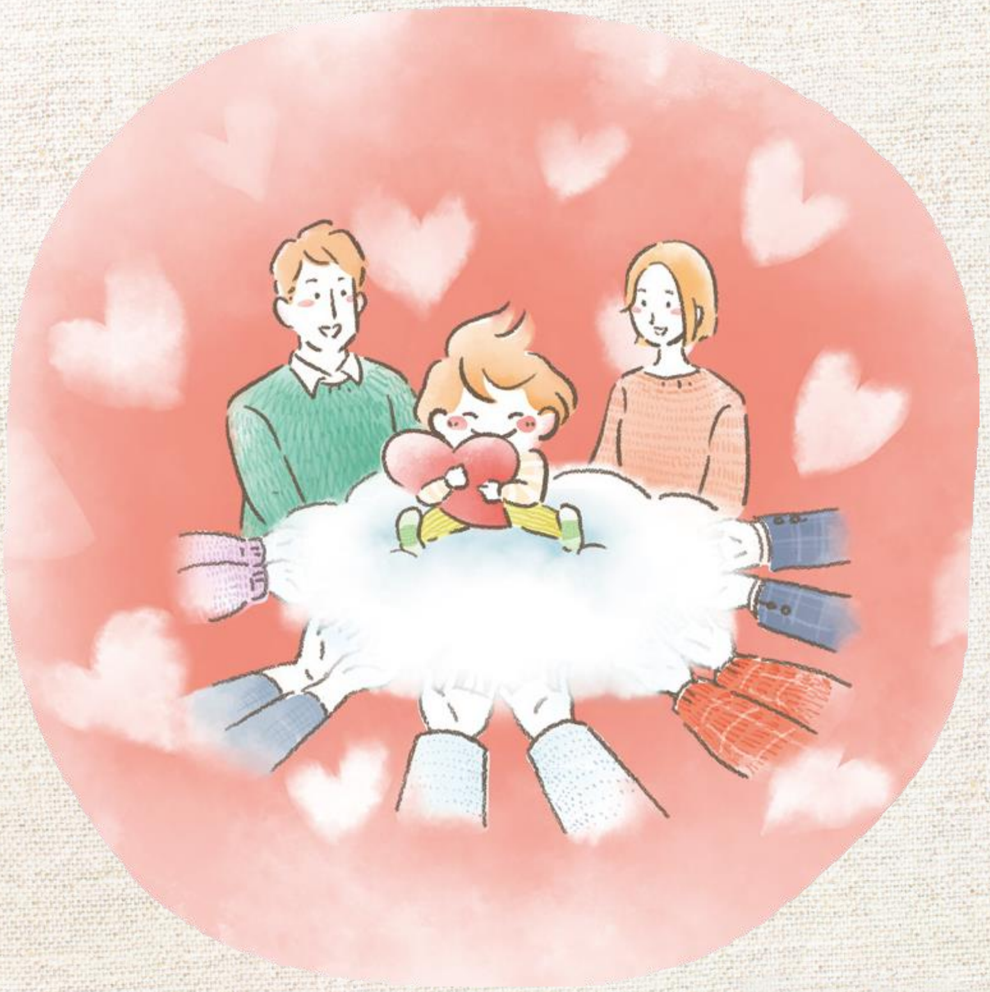
Clinical Guidance of Psychosocial Support for 22q11DS in the Transitional Period of Life

〜Essence for school and medical professionals involved
in the important life stages of child and adolescent development〜



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1 About the Guidance

With the development of medicine, more and more children are living with multiple diseases and requiring long-term medical treatment and care. While the children with special healthcare needs are now able to live in the community, the conventional medical and educational systems have highlighted the difficulty in providing services that meet the needs of the individuals and family, especially those who have multiple disabilities.

This booklet is a concise version of the guidance developed in supporting the integrated lives and well-being of individuals with 22q11.2 deletion syndrome (22q11DS) and their families. It focuses particularly on school children and adolescence, when it is difficult to provide a seamless transition of services, and describes the knowledge and attitudes that schools and health care professionals should implement.

We hope that this booklet will help those involved in education and healthcare to recognize the invisible difficulties and needs due to the overlap of disabilities in 22q11DS, which would provide a clue to realize a society that no individuals with the syndrome and their families left behind.

2 10 essential shared capabilities for those involved in support

1 Can provide a safe and secure place to stay.

Assuring that people with 22q11DS and their families can feel safe and secure is a prerequisite for support. It is necessary to rethink the nature of environments such as hospital waiting rooms, school classrooms, and workplaces, and to create places that would include people with diverse characteristics.

2 Explore and suggest specific ways to improve accessibility to support.

It is necessary to eliminate gaps in accessibility to enable appropriate use of necessary support services, taking into consideration the characteristics of the individual, such as high trait anxiety and hypersensitivity. It is desirable to propose multiple concrete options in a flexible manner, such as providing a quiet place to spend time or enabling online consultation.

3 Listen to and be close to the experience of the various complex difficulties caused by overlapping diseases and disabilities.

It is important to listen to the experience of compounded difficulties, understanding that the difficulties experienced by the individual and family are magnified by the many overlapping diseases and disabilities. If the existing support is not appropriate for a person with multiple disorders and disabilities, it is necessary to try to provide individualized and flexible support.

4 Respect diverse ways of life without imposing a uniform way of thinking or values.

We need to respect diverse values and ways of life without unilaterally imposing existing norms such as establishment of independence from caregivers. We need to be aware that the norms inherent in society and support structures themselves can be stressful for individuals and families with overlapping disorders and multiple difficulties, and we need to promote positive efforts to eliminate them.

5 Learn from the history and experiences of the individual and his/her family and respect his/her own decision-making.

It is necessary to learn from the history and experiences of the individual and family members who have lived with various difficulties and devised their own solutions, and to respect their own decision-making based on an understanding of the context. In order to support decision-making, it is necessary to provide multiple concrete options and to guarantee experiences.

6 Through dialogue, the respective needs of the individual and family can be clarified and support can be provided accordingly.

Because of experiences of not being able to obtain the support they sought, the individuals and families may have given up on the support they originally needed, or they may not know what kind of support to seek. Therefore, it is necessary to clarify the values and apparent/latent needs of the individual and family through dialogue, and to provide support accordingly.

7 Be aware of challenges within the existing support structure, including existing support practices and systems, and take positive steps to overcome them.

The overlap of disorders and disabilities can make it difficult to match the difficulties faced by the individual with the existing regular support structures, and can in fact intensify the difficulties. From the perspective of the social model of disability, we need to look at such disabilities on the side of society and devise specific ways to resolve issues on the side of support structures.

8 Communicate necessary and sufficient information in an easy-to-understand and organized manner.

Organize information about the 22q11.2 deletion syndrome and available support systems in a way that is easy for the person/family to understand and use, after the supporters has a better understanding of the syndrome and the support systems available to them.

9 Continue to learn about areas outside their own expertise and provide integrated care.

Recognize that the stove-piped approach to consultation and support by specialty does not fully address the complex problems faced by individuals and families. In order to provide integrated care, we must be willing to learn from the voices of individuals and families and to broaden our knowledge of fields outside of our specialty.

10 Build multidisciplinary and multi-agency partnerships and work together in a positive manner, confronting tensions between the parties involved when they arise due to differing interests and goals.

The more overlapping disorders and disabilities there are, the more difficult it is for a single staff to provide all specialized care. Based on this premise, build partnerships with multiple professions and institutions, and strive to provide integrated care while sharing information by holding care meetings that include the individual and family.

3 Comorbidities of 22q11.2 deletion syndrome

3-1. Heart disease

Congenital heart disease is a significant condition associated with 22q11DS. Common cardiac problems include Tetralogy of Fallot, aortic arch transection, ventricular septal defect, and atrial septal defect. These conditions often necessitate major surgery during childhood and may require reoperation in later life. Many families describe the psychological turmoil they experienced upon learning of their child’s condition shortly after birth as "like a bolt from the blue."

3-2. Ear, nose, and throat (ENT), oral surgery, dysarthria, etc.

Otolaryngological and oral surgical disorders such as cleft palate, soft palate, submucosal cleft palate, cleft lip, and nasopharyngeal dysarthria are common in 22q11DS. While an obvious cleft palate is easily diagnosed soon after birth, a submucosal cleft palate may be difficult to recognize until early childhood or later. Speech therapy is often required for dysarthria. The presence of dysarthria can cause communication stress and low self-esteem in peer groups.

Some individuals may experience recurrent epistaxis, which can be a hindrance in their daily lives.

3-3. Immunodeficiency

Cellular immunodeficiency associated with thymic hypoplasia may be seen in 22q11DS. This condition can lead to recurrent infections, which may trigger the diagnosis. Immunodeficiency and recurrent infections are often major factors preventing social adaptation. They are particularly vulnerable to the behavioral restrictions imposed by the recent COVID-19 pandemic.

3-4. Developmental disabilities and psychiatric disorders

Autism Spectrum Disorders (ASD)

ASD is often diagnosed in childhood: among those with 22q11DS, 13% were diagnosed between the ages of 6-12, 27% between 13-17, and 16% over the age of 18, according to a report.

Attention Deficit/Hyperactivity Disorder (ADHD)

ADHD is often diagnosed in childhood; among those with 22q11DS, 37% were diagnosed between the ages of 6-12 years, 24% between 13-17, and 16% over the age of 18, according to a report.

Schizophrenia, psychotic symptoms

Individuals with 22q11DS may experience schizophrenia-like psychotic symptoms such as hallucinations and delusions during adolescence and young adulthood. The prevalebnce of schizophrenia spectrum disorders is estimated to be about 20-25% in 22q11DS.

Anxiety (Anxiety Disorders)

Individuals with 22q11DS have a high rate of anxiety (anxiety disorders), 35% reported in children and adolescents. High levels of trait anxiety are associated with poor adaptive functioning and interfere with daily living skills.

3-5. Intellectual disability and cognitive developmental characteristics

Intellectual disability

The estimated prevalence of intellectual disability, including mild to severe, is 45%, and the average overall IQ for individuals aged 6-24 with 22q11DS is 71. In particular, visual-spatial cognition, arithmetic, and abstract reasoning are considered areas of weakness.

Arithmetic

Although individuals with 22q11DS can quickly read numbers and perform basic calculations, they often struggle with mental arithmetic and mathematical reasoning. These difficulties may stem from poor memorization, working memory, and visuospatial cognitive function. Therefore, it is important to establish basic arithmetic skills through step-by-step procedures.

Understanding abstract concepts

The child has difficulty in understanding abstract concepts, and delays in learning may become noticeable after the 3rd or 4th grade of elementary school, when the content of subject learning becomes more abstract. Because of their relatively good language comprehension, it is sometimes difficult for teachers to understand the child's difficulty in thinking logically based on visual information.

Memory

Memory seems to be both a strength and a weakness. While their ability to recall newly learned content after some time is age-appropriate, they struggle with recalling story details, remembering visual spaces and faces, and not only retaining information but also processing it.

Language Development

Many individuals with 22q11DS experience delays in language development. Two factors are believed to contribute to the delay: one is abnormalities of the palate and the other is cognitive developmental problems. The effects of dysarthria must be taken into account when evaluating autism spectrum disorders and cognitive developmental delays based on early childhood speech delays. Because of their difficulty in understanding abstract concepts, they may have difficulty adapting to discussion or lecture-style classes as they progress through the grades, and their ability to express themselves verbally may be underestimated by others.

Communication

Behind the obstacles in communication is a complex combination of factors such as limited vocabulary, difficulty in understanding metaphors and abstract concepts, and difficulty in understanding the intentions of others. These qualities also vary greatly from person to person, but communication can be facilitated by finding and working on the individual's strengths, such as having better input than language output.

High level of anxiety and fear

Although many individuals are highly anxious, it is easy for schools and other institutions to treat them as if they were normal children with anxiety and that they will be fine as long as they are accustomed to it. However, anxiety in 22q11DS is considered to be a characteristic of the disease. Pedagogy such as accustoming the child to a certain level of fear may be counterproductive and may eventually lead to traumatic experiences. Therefore, even if you as an educator think it is in the child's best interest, you should carefully consult with the child's caregivers and health care providers to make a prudent decision.

Hearing Sensitivity

As with anxiety and fear, auditory hypersensitivity is also likely to be a characteristic of the disorder. Responses such as accustoming the child to it may be counterproductive and, in turn, lead to traumatic experiences. In addition, we often hear that it is difficult for hospital and school staff to understand and ask for consideration when a person has auditory sensitivity to certain voices and sounds, such as children's voices (because it is common knowledge that these sounds should be pleasant for the majority of people).

Easiness to get tired (fatigue)

One of the most notable characteristics of people with 22q11DS syndrome is tiredness or fatigability which can lead to reluctance to participate in events, go to school, and other activities. Some people say that it is difficult to ask for consideration from school teachers, etc., because it is easy to be perceived as selfishness, parental spoiling, or that it is better to train the child for his/her own growth. It is necessary for those around the child to understand and watch over him or her, as the reason why he or she gets tired easily is that he or she is physically prone to cardiac and endocrine abnormalities, and mentally he or she tends to feel depressed.

4

Support in medical care

Children with 22q11DS may require medical treatment such as surgery for cardiac diseases from an early age.

In such cases, caregivers are often required frequent hospital visits, feeling the threat of their children's lives. They are often asked by medical professionals to raise the child under their protection.

Some cases are diagnosed with 22q11DS, as a result of repeated hospitalization due to infections and thus being suspected of immunodeficiency.

Even when their physical illnesses have settled and they have grown into adolescence and youth (the time when transition to adult medical care should be considered), they often have difficulty understanding the issues that may arise in various situations such as schooling, employment, marriage, and childbirth, and living independently with the necessary support.

In addition to physical disabilities from early childhood, the caregiver may notice intellectual and developmental disabilities and schizophrenia-like symptoms through school age and adolescence, and it is not easy for the person to understand and make independent decisions about the support he or she needs, including medical care.

This chapter describes the medical difficulties people with 22q11DS and their caregivers experience and the types of support needed.

Medical professionals should continue to learn about 22q and strive to provide comprehensive medical care.

Caregivers’voices

A local physician told me, "I have no experience treating this disease and don't know what to do, so please go to another hospital."

In the past, when parents explained the illness to staff, the staff would look unhappy and often took the attitude that it was difficult to provide special care. I am grateful that the current staff are willing to accept my child.

Lack of knowledge of medical professionals about 22q11DS can lead to delayed diagnosis. 22q11DS has a highly individual course and multiple diseases and symptoms coexist, so some medical professionals feel reluctant to treat them. This makes it difficult for some patients to find a hospital where they can receive treatment. Some people attend multiple departments, such as cardiac surgery, pediatrics, otolaryngology, oral surgery, internal medicine, orthopedics.... Physicians need to have an integrated understanding of symptoms in their own specialty and in other medical departments. Of course, it is impossible for a one physician alone to diagnose, treat, or provide care outside of his or her field of expertise. They need to refer patients to appropriate specialists or specialized institutions. Physicians are expected to act responsibly. For example, physicians should not simply refuse to see a patient, but should examine the patient and then appropriately refer the patient to another medical institution.

Patients who visit multiple departments may not be able to strictly follow the physician's instructions depending on the progress of treatment in each department. Accompanying children to medical appointments and coordinating each treatment is a burden on caregivers. Some people go to multiple medical departments at a general hospital, but it can be inconvenient to visit a general hospital for minor illnesses such as a cold. Medical institutions need to work together to share information and understand the treatment process and stage of each department. Physicians also need to carefully consider the burden of visiting multiple medical institutions and departments, and of diagnosing and treating multiple diseases simultaneously.

Considering the burden of having multiple diseases, medical institutions should collaborate with each other to share information on the treatment plan of each department.

Caregivers'voices

The physician decided to postpone the heart surgery because the tooth decay treatment had not been completed. The cardiac surgeon criticized the mother, saying, "Why didn't you arrange for the dental treatment to be completed before the surgery?"

Dr. A said, "Please ask Dr. B why he said that,“, and I was confused. I got tired of continuing to be a “messenger”.

Even when I consult my physician about non-specialty or social services, he always tells me to "find a hospital yourself" or "go to the government office yourself and ask.” I'm tired of playing the role of a "private care manager."

Patients and their families strive to adapt to changes in social life during each stage of infancy, childhood, and adolescence. Medical professionals need to understand the background of the patient's and their family's lives and provide treatment. Medical professionals will approach treatment at that point, but patients and their families will have to live with the illness for the rest of their lives. Medical professionals need to follow up to ensure that caregivers and siblings also live healthy lives so that families can support each other.

Consider patients’ and their families' life backgrounds and strive to provide them with thoughtful and flexible medical care.

Caregivers’voices

When my child was hospitalized, I had to attend to him 24 hours a day. I left my small child at home to be with him.

When my child was young, it was difficult for me to accompany his medical visits, leaving my small child at home.

COLUMN

Plain Japanese

According to a 2004 survey by the National Institute for Japanese Language and Linguistics, over 80% of the public expressed a desire for doctors to rephrase or add words to their explanations to make them easier to understand. In a 2008 survey, less than 20% of the public was familiar with terms such as "remission" and "QOL," and less than 40% correctly understood terms like "collagen disease" and "sepsis.“

For patients to make informed medical decisions, they must understand the meaning and content of these terms. Therefore, it is the responsibility of medical professionals to provide easily understandable explanations. Medical care providers must find ways to translate complex language into simpler terms for better patient comprehension.

When patients cannot understand the language used by medical professionals, it is difficult for them to make well-informed decisions about their healthcare. Hence, medical professionals need to communicate in a manner that is easy for non-experts to understand. By making an effort to communicate clearly, medical professionals can motivate patients to actively seek understanding. Sharing information and fostering mutual trust between medical professionals and patients are fundamental to ensuring medical safety and security.

5 Support in Education

In 1994, UNESCO proposed the Salamanca Statement to promote inclusive education for all children, including those with special educational needs. The statement emphasizes that "every child has a fundamental right to education and must be given the opportunity to achieve and maintain an acceptable level of learning." Since then, inclusive education has become an international trend.

In Japan, the Ministry of Education, Culture, Sports, Science and Technology (MEXT) has embraced this trend and compiled the "Promotion of Special Support Education for Building an Inclusive Education System Toward the Formation of a Coexisting Society." However, there is debate over the concept of inclusivity. MEXT states that in inclusive education, it is important to pursue learning together in the same place and to develop a diverse and flexible system that can provide instruction that meets individual educational needs. They believe it is necessary to prepare a continuum of "diverse learning places" such as regular classes in elementary and junior high schools, instruction through day classes, special-needs classes, and special-needs schools, and take measures to separate places. However, the United Nations review of the Convention on the Rights of Persons with Disabilities in Japan, held in Geneva in the summer of 2022, raised concerns that special needs education in separate places may not be inclusive.

Currently, children with 22q11DS in Japan receive their education within different frameworks, such as regular classes, special support services in resource rooms, special support classes, or special support education schools. However, due to the complexity and individual nature of the disease, it is challenging to initiate and continue educational guidance tailored to individual characteristics before schooling. The burden on caregivers in providing their children with a suitable education, including gathering information and choosing the right school and class to attend, needs to be addressed in the future. Additionally, many children with 22q11DS require attention to various physical symptoms, such as cardiac diseases, during school. Study delays can also be an issue if they attend treatment frequently. Many children have comorbid intellectual disabilities and neurodevelopmental disorders, which may make it difficult for them to find their school place. Their inability to fit in with the group can be traumatic. While care must be taken to ensure that excessive stress in school life does not lead to the onset or worsening of psychiatric symptoms, there are many difficulties experienced by the children and their families in education. This includes difficulties in being understood by teachers, classmates, and other parents. With this background in mind, we will summarize the support required in the education of children with 22q11DS from several perspectives.

5-1. Understanding the difficulties associated with hard-to-see disabilities and multiple disabilities

It is important to understand that even with the same 22q11DS, each individual may have very different difficulties due to physical, intellectual, and mental disabilities. Among the comorbidities described in Section 3-5, certain characteristics such as high anxiety/fear, auditory hypersensitivity, and fatigue can lead to reluctance to attend school and conflict with the school's values. This can be counterproductive and potentially traumatic for the child if they are forced to go too far. It is important to keep in mind (3) the ability to listen to and be close to the experience of various complex difficulties caused by overlapping diseases and disabilities and (4) the ability to respect diverse ways of life without imposing one way of thinking or value system in the "10 basics required of people involved in support.“ It requires careful discussion with the child, caregiver, and medical provider, and a flexible attitude that in some cases there may be an option not to attend school or not to allow the child to attend school.

Some children with 22q11DS have multiple comorbidities, and some have severe disabilities, while others have borderline disabilities with no one disease or disability being severe. When physical and intellectual disabilities are combined, there may be biased accommodation to either disability. For example, this can lead to insufficient attention to intellectual disabilities even when attention is given to physical disabilities, or insufficient attention to motor limitations due to internal disorders even when attention is given to intellectual disabilities. Even if there are overlapping disabilities, individuals with borderline physical and intellectual disabilities can be evaluated as something the individual should be able to do with effort and be required more than they are willing to put in at their own pace. Many caregivers believe that due to the borderline nature of these conditions, their child does not fit into the existing framework of regular or special needs classrooms.

Additionally, children with 22q11.2 deletion syndrome often have to attend various hospital departments in between their schooling, which makes it challenging for them to keep up with their classes.

To address the obstacles faced by individuals with these overlapping diseases and disabilities in the current educational environment, which is primarily designed with stereotypical disability characteristics, it is crucial to provide personalized support and make necessary adjustments.

Caregivers'voices

Please understand that physical, intellectual, and mental disabilities vary greatly from child to child and that there are individual differences.

Please understand that in school life, even if they seem capable on the surface, they are often physically and mentally tired and struggle to perform at their best.

In the special needs class, there are children with various characteristics. For my child, who is sensitive and vulnerable to stress, the voices in the classroom may seem loud, and being with classmates can cause a great deal of stress. Although we enrolled in the school after discussing their situation, it is very difficult for us to receive consideration for our individual physical condition and characteristics in a special needs class that has many students.

My child received consideration for exercise limitations due to his cardiac condition, but his mild intellectual and emotional difficulties were not as well understood (e.g., extreme anxiety).

The teachers at the school listened to us parents with a teachable attitude and were flexible and resourceful in their involvement that suited his characteristics.

When my child was unable to attend school, the teachers were eager to encourage attendance. However, we ultimately decided not to send them to school because we had heard from other families who regretted forcing their child to go. We are now grateful to the older families who shared those painful experiences with us.

My child has borderline physical and neurodevelopmental disabilities. Compared to children with more severe physical disabilities or children with neurodevelopmental disabilities plus intellectual disabilities, these disabilities may seem minor and one might expect them to be able to do more. However, despite being halfway there, there are still overlapping difficulties. I believe that teachers do not fully grasp the concept of "multiplication of disabilities." I would like them to understand that even if each disability is borderline, the combination of multiple disabilities can make life more challenging.

My child has an intellectual disability and a cardiac condition. After considering the divisions between intellectual and physical disabilities, he was enrolled in a special needs school for intellectual disabilities. In elementary school, my child was able to participate in moderate excursions and activities based on his physical condition and mobility limitations. However, when he transitioned to a special needs school, he struggled to keep up with his peers physically and faced difficulties in having them accommodate his physical condition. The educational setting primarily focused on properly directing energy.

5-2. Support and guidance based on needs

It is essential to listen to the voices of both the individual and their caregivers and to collaboratively develop flexible support and guidance plans tailored to each person's unique characteristics and needs, while also adjusting the environment accordingly.

Caregivers'voices

I was uncertain whether to enroll my child in a special support class or a regular class with occasional support. Although I was concerned he might struggle academically, I wanted him to experience school as a small society.

It would be good if there is an environment where they can be taught patiently and repeatedly so that they can understand.

Having knowledgeable supporters or key persons within the school who can think through issues with us would be reassuring for both the family and the individual.

Math is particularly difficult for her. It seems she doesn't quite grasp the concept of the number 2.

I hope the school values each child's individuality and offers education that enhances their strengths based on their interests, teaches daily life rules, and provides experiences in communication and interaction.

I want them to understand that although they seem to be able to do everything, they get tired easily and are mentally delicate and sensitive. Please treat them normally, but understand that there are many things they cannot do, and I don't want them to be pushed beyond their limits.

When my child stopped attending school, no one listened to how tough school life was for him. The principal even suggested, "Isn't the problem with your child?"

From the voices of caregivers, it is clear that children with 22q11DS face complex and highly individualized challenges due to overlapping conditions. Each child has a wide range of characteristics and needs, requiring more flexible support tailored to their physical and mental state at any given time. When developing individualized education and instructional plans, it is essential to gather information not only from medical professionals but also to listen to the wishes and hopes of the child and their caregivers. Observing what the child can do, or is close to achieving, and focusing on their interests and concerns is crucial for creating specific support and instructional goals. Additionally, sharing information with the child, caregivers, and relevant parties is vital to identify the necessary support during academic learning, adjustments for sensory sensitivities (such as sounds and smells), communication with friends, and support in social situations (including individual instruction). Specific strategies should be outlined for individualized education and instructional plans.

For parents sending their child to school for the first time, they may not have sufficient information about the school or education system. The burden of gathering information and considering an educational environment that can flexibly address intellectual and physical challenges should also be acknowledged. Additionally, as children progress through grades and face difficulties adapting, parents are sometimes blamed for making school choices. It's important that experts thoroughly evaluate the child's characteristics and the suitable educational environment, share this information, and collaboratively consider the appropriate education with the caregivers.

5-3. Support focused on the strengths and advantages of the individual

People with 22q11DS exhibit a wide range of individual differences, and have many strengths and advantages. In addition to assessing their disabilities and needs as discussed in sections 5-1 and 5-2, it is equally important to focus on their strengths. It is the individual and their caregivers who know the individual's strengths best, so it is beneficial to ask them for insights.

Commonly observed strengths include:

- Memorized calculations using given formulas
- Reading and understanding basic information
- Spelling and grammar
- Memorization
- Focus on simple tasks
- Computer skills
- Typing speed
- Motor-sensory skills (e.g., dancing)
- Rhythmic and musical talent
- Willingness to learn
- Pleasant personality
- Kindness

5-4. Establishment of a cooperative support system

It is necessary to establish a support system in cooperation with medical care, welfare, and government agencies.

Caregivers'voices

Not knowing which school was the best, we had to research each one individually and gather information.

We would like to have someone who can connect us in cooperation, rather than the foster caregivers bridging all the gaps.

I am raising my child with independence in mind, but I don't know how to create a place for him or her to stay after high school.

I would like to see information shared with caregivers and collaboration with medical and social services regarding understanding of the disease. We would like to know what kind of information the caregivers should tell the school and they should be able to share with the school.

It would be reassuring for families if there is an environment where medical professionals, school personnel, and others understand the disease and can bridge the gap using pamphlets and tools created by family associations and medical institutions.

Another difficulty for families in education is the lack of cooperation between schools and other institutions. This is due to the current situation, as mentioned by the caregivers above, where families have to pass on information to each place by themselves to bridge the gap. For families who are involved with many medical institutions and support organizations, it is a big burden for families to grasp and convey all of this information. For this reason, there is a need for places and environments where schools and families, education and medical care, education and welfare, or all of them can collaborate, and for people who can connect them. In addition, the support provided by municipal boards of education in consultation services when making school choices, and the existence of partnerships between schools and other services such as after-school day care services, can be a source of strength for the family.

Medical professionals can help make mental difficulties, such as hypersensitivity and anxiety, more understandable by documenting them in medical certificates. This can help in obtaining support and providing clear feedback to educational institutions based on psychological assessments. It has been reported that children with 22q11DS tend to have higher expressive ability than cognitive ability. Because of their expressive abilities, those around them tend to estimate their abilities as high, but in reality, they often lack proficiency in life skills that are essential for independence. Another prominent characteristic of children with 22q11DS is their difficulty with arithmetic. It would be good if information on these characteristics could be shared between the medical and school settings, and supportive measures could be taken.

Considering education for children with 22q11DS who do not fit into the existing educational system because of the difficulties associated with their multiple disabilities may provide a new perspective on the educational system for all children, not just those with special needs.

COLUMN

How School Teachers Can Help

Unfortunately, the name of the disease, 22q11DS, is still not well known. In many cases, school teachers have never heard of the disease before and may only learn of it when they meet a child who has actually been diagnosed with the disease. We would like to share with teachers who have picked up this booklet in their search for better ways to support their children.

1 Tell the child and caregivers that you know the name of the disease 22q11DS, or that you have never heard of it before but would like to discuss it together.

Children and caregivers often talk about their hurtful experiences of being told "I don't know" or "I don't understand" in various situations and not being able to discuss the issue as much as they would like. Conversely, it is a great relief to be accepted as "knowing the name of the disease" or "you have such a diagnosis. Establishing a relationship where parents, children, and schools can feel comfortable consulting with each other will be the first step in providing support.

2 Syptoms and severity vary greatly from person to person, so please pay attention to the characteristics of "this child" in front of you.

The mmanifestation of symptoms and stumbling blocks in children with 22q11DS is characterized by large individual differences. As noted earlier in this guidance, there can be a variety of comorbidities, but how many of these diagnoses a person receives and the extent to which they cause stumbling blocks in daily life varies from person to person. I believe that this diversity and individual differences are the main reasons why teachers have trouble dealing with this issue at school. First of all, we would appreciate it if you could confirm with the individual, his/her caregivers, and in some cases, medical institutions and other related parties, what kind of illnesses or difficulties "this child" has when he/she comes to school.

3 Many children with 22q11.2 deletion syndrome seem to be easily anxious. A safe and secure environment that respects the child's pace and allows the child to say "help" when in need is desired.

It is also said that children with 22q11DS tend to be better at verbal interactions than at understanding things, and may be perceived by others as "understanding" even if they do not actually understand. As a result, they tend to demand higher standards from those around them. Please be aware that children who try their best to meet expectations may, inwardly, feel constantly anxious because there are so many things they don' t understand. Children who try too hard and become tired, or who want to escape from anxiety, leading to unusual behavior, need to be involved in a way that does not require them to try too hard. Respect the child's pace and watch his or her efforts. In doing so, it is helpful to discover the child's strengths and interests. Some children with 22q11DS excel in the arts, such as music and painting, and being recognized for their strengths can boost their self-esteem. An environment in which the entire class recognizes each other and can say "help" when in need is the foundation for a safe and secure school life for the children.

4 Teachers may also feel difficulties in dealing with the situation and may want to discuss it with someone. At that time, please consider consulting with related organizations together as well.

It is likely that teachers will often find it difficult to cope with a child in front of them who has an unfamiliar disease name and needs support. In such cases, they may consider using a consultation system that includes administrators and others in the school, and consulting with a school counselor or a school social worker. In addition, the individual is often involved with other parties outside of the school, such as a hospital. We encourage you to contact these non-school parties when they are available. By sharing your ideas with them and discussing any difficulties you may be experiencing, you may be able to find better ideas and reduce the burden of having to deal with the situation on your own. Teachers also need a safe and secure environment.

6 Support for transitions (from childhood to adulthood)

6-1. Transitional support

Although children's cognitive development is still in its infancy and their self-determination may not be sufficient, medical treatment should not prevent children from fully recognizing their own situation and making their own future decisions as much as possible.

To support the continuity of medical care and life for children with illnesses and promote their autonomy, continuous childcare support from an early stage, medical care support considering developmental diversity, and active regional cooperation (including welfare and education) are more important than ever.

1. Strengthen the recognition of medical staff
→ Establish a system (especially pediatricians) to recognize psychosocial problems of children with illnesses.

2. Strengthening of the psychosocial support system
→ Establishment of a support system involving multiple professions such as liaison-consultation psychiatrists (child mental health specialists), psychologists, social workers, etc.

3. Implement regional cooperation (welfare, education, medical care) from the early stage (e.g., use of the system for specified chronic pediatric diseases, connection to public health nurses in charge, etc.).

Caregivers'voices

（There are (caregiver +child)/2 adolescent challenges.

After graduating from pediatrics, I transferred to internal medicine, but as an adult, I had different departments to see depending on my symptoms, so I didn't have a doctor who had a comprehensive knowledge of my condition, so I had to run to the pediatrician for emergencies.

6-2. Disease notification

In some cases, the diagnosis is informed soon after the child is born, while in others it may take a long time before the diagnosis is given. Some caregivers feel various sensations of hopelessness, helplessness, and guilt when their child is diagnosed, so it is necessary to communicate this with psychological considerations in mind. They also feel helpless and frustrated by having to wait years before their child is diagnosed.

Caregivers'voices

I was noticed of my child's diagnosis when I had just given birth to my child. I did not want to believe it.

I don't know when or how old to make the notification, who should do it, or how much to tell my child.

I thought that disease notification was necessary to understand that having various physical ailments and being uncomfortable and in pain with things that are normal and okay for many people are due to the 22q11DS.

COLUMN

18-year-old barrier

In transitions, the problem of the so-called "18-year-old barrier" arises because of the division in welfare between services for children under 18 and services for adults over 18. In the field of medical care, some pediatricians and child psychiatrists operate bylaws that uniformly refer patients to other hospitals when they turn 18 years old. This has a particularly large impact on people like those with this syndrome, who often have multiple disabilities and mental illnesses after adolescence and need seamless support from multiple professions and pediatric and adult services. The impact of the 18-year old barrier is immeasurable for those individuals and families who are at a crossroads in their lives, whether to pursue further higher education or to find employment. It is desirable to have seamless government services and local case managers, but for the time being, it would be realistic to focus on realizing seamless support by having doctors, clinical psychologists, and social workers at medical institutions act as a bridge and holding staff meetings.

7 Family support

7-1. Support for caregivers

Caregivers of children with illnesses or disabilities experience a variety of physical and emotional burdens. 22q11DS is a life-long condition with changing symptoms that can place a long-term psychological burden on caregivers and cause a wide range of difficulties. Caregivers of children with 22q11DS experience a variety of psychological difficulties, including shock at the child's diagnosis, separation from the child due to treatment, and fear of the child's death. The amount of care that is regularly imposed on caregivers during the course of parenting is enormous, and many caregivers feel overwhelmed by this. Cardiac diseases, circulatory symptoms, and frequent infections that are discovered shortly after birth leave the caregiver with no time to rest and very few opportunities for relaxation. This burden of care takes a toll on the caregiver's life. It has been shown that caregivers must limit their own work to care for their children and tend to isolate themselves from society by devoting their lives to child rearing. Because few professionals in either the medical, educational, or social services have a thorough understanding of 22q11DS, it tends to be the role of the caregiver to communicate and coordinate between support agencies in order to receive appropriate support. The burden on the caregiver is further increased when support is not adequately provided. The caregiver feels overwhelmed in his or her role as a surrogate, educator, and coordinator of services. The inability to care for children as the caregivers get old is one of the most serious concerns. It is imperative that society bear as much of the role that the caregiver assumes as possible, and that a social system be established to support the individual's disability. Thus, it is important to note that the burden on caregivers is amplified not only by dealing with the symptoms presented by the child, but also by the challenges on the part of the system.

With this understanding of the difficulties experienced by caregivers, it is essential to expand public support. On the other hand, of course, it is also true that there are many caregivers who speak positively about their experiences raising children with 22q11DS. It is most desirable to build a system in which society as a whole supports children living with diverse diseases, rather than only the caregivers.

Caregivers'voices

Every day I worried that if I took my eyes off him, he would die. When I was left alone with my child by the nurse, I remembered how I used to spend my time alone.

I'm not a messenger pigeon (of doctors in department A and B). I always thought, "I am the one who needs help".

My child with 22q11.2 deletion syndrome couldn't keep up with his schoolwork at all, so we , the whole family came together to help his learning.

I am worried about my child's future. I want her to be able to live on her own without relying on her siblings. I always wonder if she will be able to survive on her own.

7-2. Siblings

When a child is ill or disabled, caregivers devote much of their time in life to the child's treatment and care. This is said to reduce the attention of the caregiver to the "siblings" (Kyodai-Ji). It has been shown that siblings can be jealous for the individual with 22q11DS, but they also know that the illness or disability is not the fault of the caregiver or the individual, so they are more likely to feel guilt and shame about their jealousy. Siblings gradually learn to repress their feelings and avoid stressful situations, and they rationally accept the diagnosis of illness or disability and its associated limitations. Siblings may also take on a supportive role for their caregivers who devote their time to treatment and care. For this reason, siblings tend to be patient and empathetic. Some siblings are aware of their own role as caregivers when their caregivers become elderly. Siblings who have learned to behave rationally in their environment may seem at first glance to be living a stress-free life. However, social support must be provided, paying attention to the development of the sibling himself/herself as well as the individual with the illness or disability.

Of course, families and siblings feel differently, and not everyone experiences this way. However, it is important for supporters to understand that siblings of individual with illnesses or disabilities may have difficulties that are not well known and difficult to express in words. Support for siblings is still insufficient, and further development is desired.

Caregivers'voices

I feel that I want my child with 22q11DS to grow up on his own, without bothering his siblings.

Siblings' voices

Somewhere I have the feeling that if something happens to my parents, I will be responsible for it.

From a young age, my role was to support the parents who were busy taking care of my brother with 22q11DS.

22q11DS Special Clinic, The University of Tokyo Hospital

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