

How can we support the parents of children with a life-shortening disease?

When a child is diagnosed with a life-shortening disease, this has a profound effect on the entire family. How do parents react to receiving such news? What can we as health professionals do to support them when their child is diagnosed with a rare progressive degenerative condition?

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A disorder is regarded as rare when it affects fewer than 500 of the general population (1:10 000). More than 30 000 people in Norway live with a rare disorder, many of whom are children. Several of these diseases show a progressive course where the child develops normally early in life. These diseases may be muscular, for example Duchenne muscular dystrophy; neurological, for example neuronal ceroid lipofuscinosis; or metabolic, for example adrenoleukodystrophy (1).

The search for a diagnosis

Although the interval between the parents suspecting that something is wrong and the diagnosis of a rare disorder may be short and brutal, it can also be long and challenging. Since rare disorders are exactly that, most doctors will never have met a patient with the diagnosis in question. Often no suspicion will have been aroused, and consequently the necessary tests will not have been carried out (2). The parents may feel they are dismissed as «over-anxious parents», and a fair number say that their concerns about their child's first symptoms were disregarded (3).

Trying to establish a diagnosis can be very stressful for the parents (4), but the parents can also make helpful contributions through their searches on the internet (2). The information they find should be taken seriously, and should be included in the dialogue with the doctor (5). Clinical experience indicates that if the parents are to continue to have trust in the health care services, it is important to listen to their suspicions and to take them seriously.

In the course of the diagnostic process, the doctor may perhaps suspect a particular disorder and may decide to carry out a specific diagnostic test. It is then important to consider whether to inform the parents about the reason for conducting the test.

Often parents have not heard of the disorder prior to its diagnosis (3), but when asked, they often want information about the disorder before tests are carried out and to be able to influence when and how much information they are given. Parents say that it is crucial that they are met with empathy, hope and friendliness when such information is given. We know that the need for information and emotional support at the time of diagnosis is closely linked to the parents' prior knowledge and emotional state (6).

Communication

«They said nothing in the news about the world falling apart today» (7). How a child's diagnosis is communicated, and how the shock affects those concerned represent an event and a memory that may influence how the family copes with its new everyday life (8). It is vital that the parents get the information they want, that they understand it and that they remember it afterwards (3). However, the triggering of acute stress reactions may challenge the flow of information. The use of particular approaches may improve the handling of the situation.

Based on a review of the research in the field, Boyd (9) developed the following guidelines for delivering diagnoses in children's neurology departments: Have a meeting with the parents as soon as possible after the diagnosis has been confirmed – in a private and friendly setting. Ask both parents to be present or in the case of single parents, that they are accompanied by someone who can support them. The person in charge of the meeting should know the family and be an expert on the disorder. All attending personnel should have played a part in the diagnostic process. Communicate the diagnosis early on in the meeting, and then ask the parents about their knowledge of the disorder. Give appropriate information about the prognosis, development, treatment and services. Build hope by referring to experience and research. Listen to the parents when they

express their feelings. Give them information about relevant services they can be referred to, and mediate contact with user associations if so desired. Offer the parents a written summary of the information given during the meeting as well as the opportunity to have a further conversation at a later date. At the end of the meeting, the parents should be given the opportunity to be alone in a private room.

Health professionals have a duty to ensure that children receive good information about their diagnosis (Patients' Rights Act sections 3–4 and 3–5). An assessment should be made of whether the child should be present when the diagnosis is delivered. A doctor who is well prepared can help to establish mutual understanding and openness by communicating the diagnosis to the family together. If the child accompanies the family to the hospital but is not present during the consultation, plans for how the child is to be looked after should have been made. Parents who have to go to their child immediately after a serious diagnosis has been delivered often find this very difficult (3). It is therefore important that the department has resources available to look after the child if required.

It is essential to provide guidance for parents about how they can talk to the child (also siblings) about the disorder. This is particularly important if the child is not present when the diagnosis is delivered. Many parents are uncertain, and need advice. Children know that they have been examined by the doctor, and they often notice that their parents are upset. The first step in informing the child may therefore be to say that the doctor has found out that he/she has a disorder, and that naturally the parents are sad because of this. The most important information parents give is linked to the symptoms or restrictions the child experiences in daily life and how these are dealt with (10).

Giving information is a process in which health professionals must help the parents to make choices that are right for the family at various stages of life. There are many



Illustration: Sylvia Stølan

options between «telling all» and «keeping quiet», and most families learn as they go. If the child has a rare disorder, parents can contact the Norwegian National Advisory Unit on Rare Disorders (NKSD) for advice and guidance (11).

Training can enhance the doctor's skills in talking to parents and children about progressive disorders (12). It is vital that the department prioritises this. When a diagnosis is to be communicated, the doctor must prepare what to say and consider how their own and others' reactions can be dealt with (13). As a health professional, it is not unusual to deal with difficult emotions by giving more information (14), but the parents need a doctor who listens and shows empathy (15). In order to be able to do this, support from colleagues is needed. If the doctor suppresses the difficult emotions that arise in challenging meetings with patients, there is a greater risk of subsequent emotional exhaustion than when he/she seeks support (16).

Support for parents

The crisis reactions that parents experience at the time of diagnosis can feel frightening.

Information about normal reactions may help to prevent the extra burden of worrying about one's own reactions (17), and this is recommended in the national guidelines for crisis management (18). The doctor should focus on the significance of routines, food and sleep in everyday life going forward, and how family or friends can perhaps provide practical help. The immediate reactions of some parents will be so strong that the mental health services may need to be contacted. In my experience, the emergency team in both the primary and the specialist health services can be on standby if they receive sufficient information regarding the traumatic nature of the diagnosis.

In the further follow-up of the child, attention should be paid to the parents' adaptation. Common physical reactions include greater activity, muscle tension, sleep problems, abdominal problems and impaired immunity. Common mental reactions include intrusive memories, concentration and memory difficulties, a feeling of unreality and obsessive thought patterns. The natural course of a crisis is that individuals are partly protected at the start and that strong emotional reactions follow only

after a period of time (19). Those who had no need of psychological support and guidance at the start may well need this later on.

Anticipatory grief is a term that has been used to describe the process that the parents of a child with a shorter life expectancy embark on, and which can entail many of the same thoughts, emotions and reactions that are experienced after a death has occurred (20). For a parent, being in this situation is a major strain but parents also describe how time assumes a different value, that the experience of life becomes stronger and that the days can also be experienced as good (21).

The parents' perception of coping is keyed to the resources they have access to, but many have a high threshold for seeking help (21). Health professionals should also help provide information and advice to the network around the family, if necessary in cooperation with the centre for rare disorders that has responsibility for the diagnosis (11). Supportive dialogue provided by the primary health care services (see mental health care on the municipality's website) may provide useful assistance to parents, children and siblings. In the case

of psychological problems that affect everyday functioning, the specialist health service is the correct instance. It may be appropriate for the specialist to provide a statement about the nature of the diagnosis in their referral to the GP.

Conclusion

Taking good care of the parents is an investment in the shared future of the child and the family. Those of us who work with families who are thrust into the nightmare of their life know how wise and robust both parents and children can be. Good communication of the diagnosis, focus on the family in the follow-up, involvement of the mental health service and a sound inter-agency collaboration is crucial in supporting these families.

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