

## THE COMPETENT NOVICE

### Organ donation

Paul J Frost,<sup>1,3</sup> Stephen Leadbeatter,<sup>2</sup> Matthew P Wise<sup>3</sup>

<sup>1</sup>Division of Medical Education, Cardiff University School of Medicine, Cardiff CF14 4XN

<sup>2</sup>Department of Medical Genetics, Pathology and Haematology, University Hospital of Wales, Cardiff

<sup>3</sup>Critical Care Directorate, University Hospital of Wales

Correspondence to: P J Frost  
FrostP1@cf.ac.uk

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This series aims to help junior doctors in their daily tasks and is based on selected topics from the UK core curriculum for foundation years 1 and 2, the first two years after graduation from medical school.

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▶ Mabel Chew discusses organ donation with Paul Frost in a BMJ podcast

#### Box 1 | Organs and tissues suitable for transplantation

Organs: kidney, heart, lung, liver, pancreas, small bowel

Tissues: cornea, bone, tendon, skin, heart valve.

#### KEY POINTS

Organ and tissue transplantation can save and greatly improve lives, but a paucity of organs means that many patients die awaiting transplantation

Junior doctors' lack of familiarity with donation contributes to low donation rates, but donation should be considered as a routine part of end of life management

Specialist nurses for organ donation can be contacted through the hospital switchboard and can advise junior doctors on any aspect of donation

Families are more willing to consent to donation if they understand brain stem death and perceive that their relative has received high quality care.

Junior doctors working in general hospitals are likely to encounter organ donation from deceased donors. They can gain understanding of this complex process by accompanying senior colleagues

Organ and tissue transplantation leads to dramatic improvement in patients' quality of life and outcomes. Unfortunately, many people die while waiting for a transplant because of the paucity of available organs.<sup>1 2</sup> Aside from this human burden, the economic impact of this shortage is considerable. The annual cost of treating a patient with dialysis is £23 177 (€26 126, \$36 319) compared with an outlay of £42 025 for a renal transplant followed by £6500 a year.<sup>2</sup> Donation may provide some comfort to a grieving family and have a beneficial effect on the bereavement process.<sup>3</sup> Despite these affirmative aspects of donation, evidence suggests that medical students and junior doctors lack familiarity with this issue, contributing to low donation rates.<sup>4 5</sup>

This article focuses on donation of organs and tissues from deceased, not live, adults, circumstances that are likely to be encountered by junior doctors working in any UK general hospital. Deceased donors may be brain stem dead with a beating heart (sustained by mechanical ventilation) or asystolic (cardiac death). In the UK there were 959 deceased donors in 2009-10, of whom 624 were donors after brain stem death and 335 were donors after cardiac death (non-heart beating donors).<sup>2</sup>

All transplantable organs (box 1) can be retrieved from a brain stem dead donor. Kidneys, pancreas, liver, and lung can be recovered after cardiac death, but in either circumstance organ retrieval must be swift because ischaemic organs deteriorate rapidly. Organ retrieval is usually facilitated in the intensive care unit or, less commonly, in the emergency department.

Transplantable tissues have a low metabolic rate and, can be removed up to 48 hours after cardiac death (except for corneas—eyes must be removed within 24 hours of cardiac death). With appropriate consent, successful tissue retrieval can occur following almost any death in hospital, and junior doctors should consider raising this issue with senior colleagues when verifying such deaths.

#### How best to do it?

The clinical management of donation is complex. The best way for junior doctors to gain understanding is to accompany senior colleagues involved in the process, which in the intensive care unit usually occurs in steps.<sup>6</sup>

#### Step 1: discussions with family

The primary focus of clinicians managing critically ill and injured patients is to restore health and an acceptable quality of life. Unfortunately, about a quarter of patients in UK intensive care units are unresponsive to treatment and die.<sup>7</sup> Those with devastating head injuries or intracranial haemorrhage often become brain stem dead. In these circumstances mechanical ventilation started during treatment allows continued circulation and organ perfusion, although asystole (usually within 72 hours) is inevitable. Unsurprisingly, relatives can find the concept of brain stem death difficult to understand and accept, and careful explanation may be required (see Case scenario part 1).

When patients who are likely to die have been identified, doctors and nurses make certain that the patient's family and friends comprehend the expected outcome. Once the family understands the futility of continued aggressive treatment, discussion is then about how treatment will be withdrawn. The family is reassured that although treatment will be discontinued, care will continue to ensure that the patient remains comfortable and free from distress. A discussion of donation is not started at this time, unless the topic is raised by the family, to avoid any mistaken perception that the decision to withdraw treatment has been influenced by the need for organ donation. The issues are independent of each other; in the context of brain stem death, some evidence indicates that separating the request for organ donation from notification that the patient has died is associated with reduced rates of refusal.<sup>8</sup> Families must be allowed time in private to come to terms with the situation and time to ask questions before intensive treatments are withdrawn.

**Step 2: discussion with specialist nurse**

Once the likelihood of brain stem death or the futility of continued treatment has been conveyed to the family, the senior clinician will alert the specialist nurse for organ donation to the presence of a potential donor. This step sometimes occurs before the initial discussion with the family, as early involvement of the specialist nurse can prevent needless and potentially distressing discussion with families in circumstances where donation will not be possible. This specialist nurse is employed by the NHS Blood and Transplant service (NHSBT) and usually has a background in critical or emergency care. Most large intensive care units now have these specialist nurses based (embedded) on the unit. Their primary role is to ensure that families are given the opportunity to consider donation when a relative dies, thereby increasing the number of organs and tissues available for transplantation.<sup>9</sup> He or she can also advise junior doctors on any aspect of donation. The specialist nurse checks the potential donor's details against the NHS organ donor register—currently over 17 million people are registered as willing to donate in the event of death. The nurse also screens potential donors for contraindications to donation; the major ones are prion disease, uncontrolled systemic infection, and disseminated malignancy.<sup>10</sup> Physical condition matters more than age—successful transplants have been performed from donors in their 70s and 80s.<sup>11</sup> Full selection criteria are set out in Annex A of Directions 001/2006 and the Code of Practice on Donation issued by the Human Tissue Authority.<sup>12</sup>

**Step 3: establishing death**

Box 2 describes diagnostic criteria for brain stem death accepted by courts in England, Wales, and Northern Ireland as sufficient to determine death.<sup>13</sup> Otherwise, death is established by a period of apnoea and asystole for at least five minutes; followed by confirmation of the absence of the papillary responses to light, the corneal reflexes, and any motor response to supra-orbital pressure.<sup>13</sup>

**Step 4: discussion with the coroner**

Where a potential donor is dying, the doctor must consider whether to refer to the coroner when death occurs (box 3). If such a referral is made, consent will be required from the coroner before organs or tissues are retrieved. However, where death results from injury involving only the head, retrieval of organs from chest and abdomen often does not impede a coronial inquiry or a police investigation into a suspicious death.<sup>14</sup>

A coroner, on being approached about an individual case, might ask the clinician to liaise with a coroner's pathologist or forensic pathologist about the appropriateness of organ retrieval, in particular whether retrieval might obstruct investigation of how the person died. In our experience, organ or tissue retrieval will rarely obstruct an investigation. Department of Health guidance mentions that refusal to agree to a donation can distress donor families and strongly advises that coroners clearly explain their reasoning to transplant co-ordinators so that families can be informed.<sup>14</sup> It is not necessary for a forensic pathologist to be present at the retrieval operation, as the docu-

**CASE SCENARIO PART 1**

A 26 year old motorcyclist suffers a devastating head injury in a road traffic crash. After admission to intensive care he is unresponsive to treatment and seems to be brain stem dead.

The consultant intensivist speaks to the motorcyclist's parents and explains the nature of their son's injuries. He says that their son may already be brain stem dead and that he will perform tests to check whether this is the case.

The father asks: "What is brain stem death?"

The intensivist replies: "Brain stem death is the complete and permanent loss of function of the brain stem, the region of the brain responsible for maintaining life. Brain stem death means that the person has died."

The father says: "So if you find my son is brain stem dead, he is really dead?"

The intensivist replies "I am afraid that is the case. But we will do some tests now to check very carefully."

The father asks: "If my son is dead why does he feel warm? Why is his chest moving and his heart beating?"

The intensivist replies: "Usually when a person dies the heart and breathing stop first, depriving the brain of oxygen, so the brain then dies. However if the brain dies while a person is connected to a mechanical ventilator, oxygen is supplied to the heart, so it continues to beat, and that is why your son feels warm. Once mechanical ventilation is removed the heart will stop."

After this conversation, the intensivist leaves the family and alerts the specialist nurse for organ donation, who establishes the absence of obvious contraindications to donation. The intensivist and a consultant colleague test for brain stem death and confirm that the patient has died.

The intensivist returns to speak to the parents and says: "We've carried out the tests twice, and I am sorry to tell you they confirm that your son is brain stem dead, he has died as a result of his injuries."

The parents are inconsolable. After a period of time the father asks: "What happens now?" The intensivist asks if he might invite the specialist nurse to join them to discuss the next steps.

**CASE SCENARIO PART 2**

The specialist nurse for organ donation joins the intensivist and the motorcyclist's parents and explains her role.

The intensivist asks the parents: "Do you think that your son would have wished to be an organ donor?"

The father replies that he doesn't know and that his son had never spoken about this issue; while the

mother says that she thinks her son would want to be a donor.

The specialist nurse says that it is often the case that this matter is not discussed and acknowledges how difficult it must be for the parents to consider at this time.

The father asks: "What would the process entail?" The nurse briefly describes the necessary steps,

mentioning that sometimes donation can be comforting to bereaved families.

The intensivist says that the decision to pursue organ donation is entirely up to the family and that if they would prefer not to do this then that decision will be respected. Other relatives arrive and the family are left in private to consider the issue further.

**Box 2 | Diagnosis of brain stem death<sup>6</sup>****Preconditions**

Three preconditions must be met before testing for brain stem death can occur:

- (1) The cause of brain damage must be known
- (2) Reversible causes of coma have been excluded, including: sedative drugs, neuromuscular blocking agents, hypothermia (core temperature must be greater than 34°C at the time of testing), and circulatory, metabolic, and endocrine causes (for example profound hypotension, hypoglycaemia, hypernatraemia, and hypothyroidism)
- (3) The patient is apnoeic and mechanically ventilated.

**Clinical tests for brain stem death**

Conducted by two doctors both registered with the General Medical Council for more than five years, of whom one is a consultant and neither is part of the transplant team. The clinical tests for brain stem death must establish the following.

- (1) Absence of brain stem reflexes including: pupillary reflexes, corneal reflexes, oculo-vestibular (caloric) reflexes, cough and gag reflexes, and absence of central response to a painful stimulus
- (2) Absence of respiratory movement during the apnoea test. The patient is preoxygenated for approximately 10 minutes and PaCO<sub>2</sub> is increased to around 5 kPa by reducing the minute ventilation. The patient is then disconnected from the ventilator while oxygenation is maintained by insufflating oxygen at 6 l/min down a tracheal suction catheter passed down the endotracheal tube. Over a 10 minute period the patient is continually observed for any respiratory movements; during this time the PaCO<sub>2</sub> is allowed to exceed 6.65 kPa—once this threshold is reached the patient is reconnected to the ventilator.

**Repetition of tests**

The tests are always done twice to remove the risk of observer error and to reassure the family; there is no prescribed time interval between testing. The time of death is recorded as the time when brain stem death has been demonstrated by the first set of tests.

**Box 3 | Questions to consider before retrieval**

- Is this death to be referred to the coroner? For example, referral is required for deaths following road traffic accidents.<sup>19</sup>
- If so, does the coroner consent to organ retrieval?
- If so, does any issue exist that requires discussion with a pathologist, such as whether organ retrieval would interfere with a coronial inquiry?
- Is there any contraindication to organ or tissue retrieval, such as disseminated malignancy?
- Is there consent to retrieval?
- Did the dying person express consent during life? If such consent is present, is there any objection from family members and, if there is, can that conflict be resolved?
- If there is no consent from the dying person, does that person have a nominated representative? If there is consent from the nominated representative, is there any objection from family members and, if so, can that conflict be resolved?
- If there is no nominated representative, who in the family has the “highest qualifying relationship” and does that family member wish to consider the question of donation? (Consent must be obtained from the highest ranking relative, who may not be the next of kin. In order of precedence the highest qualifying relatives are: spouse or partner; parent or child; brother or sister; grandparent or grandchild; niece or nephew; step-father or step-mother; half-brother or half-sister; friend of long standing)
- If the person in the highest qualifying relationship consents to retrieval, is there any objection from family members lower in the hierarchy of qualifying relationships and, if so, how can that conflict be resolved? (Unanimity among family members is not strictly necessary but highly desirable)
- Is blood taken within seven days before death available for serological testing for viral infection?
- If not, had the patient received a transfusion in the 48 hours before death that might have resulted in 50% dilution of blood volume? (Tissue retrieval is generally precluded in these circumstances because viral assays may be unreliable, but organ retrieval may still proceed if the risk of death in the potential recipient without transplantation is considered greater than the risk of acquiring a viral infection from the transplanted organ)

mentation of internal injury should be within the expertise of any surgeon and clinical photographer.

**Step 5: discussion with the family**

Once the family have understood and accepted that their relative is brain stem dead, or that treatment is to be withdrawn on grounds of futility, the option of donation should be discussed with them unless obvious contraindications are present. The request for the family to consider organ donation should not be abrupt but should emerge from a caring, empathetic relationship between clinical staff and the donor's family.<sup>15</sup> Usually the senior clinician (rather than a junior doctor) and specialist nurse for organ donation adopt a collaborative approach to these discussions (case scenario part 2) but sometimes clinicians prefer to undertake these discussions independently. A recent randomised controlled trial did not show any increase in consent rates when collaborative requesting was used in place of routine requesting by the patient's clinician.<sup>16</sup>

The principal aim of this discussion with the family is to establish what would have been the patient's wishes. These may be known if the patient is on the NHS organ donor register, but otherwise the family's intimate knowledge of their relative's beliefs and character will enable them to come to a decision. Although all major religions have stated their support for the principles of organ donation and transplantation,<sup>17</sup> religious beliefs may occasionally be a barrier. For example, some orthodox Jews object to any interference with the body after death and may wish to consult a rabbinic authority before consenting to donation. Informed consent to donation must be sought and documented in compliance with the Human Tissue Act 2004 and the Directions and Codes of Practice issued by the Human Tissue Authority.<sup>18</sup> Families must never feel under any pressure to agree to donation, and must be assured that their decision will be respected. If organ donation is refused, treatment is withdrawn according to the unit's usual policy. If the family agrees to donation then the specialist nurse will undertake a donor risk assessment, including enquiring about behaviour (such as intravenous drug use) that may prevent donation.

The family will be asked if they have any requirements for laying out the body and told that they will still be able to see the deceased after the retrieval procedure. The questions for consideration by the specialist nurse for organ donation are summarised in box 3.

In controlled donation after cardiac death retrieval of organs is usually started within five minutes of the diagnosis of death so as to minimise the warm ischaemia time. This means that families only have a brief period with their deceased relative. If they require more time the donation process may need to be stopped entirely.

**Step 6: organ preservation, allocation, and retrieval**

In brain stem dead donors, adverse pathophysiological changes may threaten the viability of transplantable organs and may require specific approaches—for example, inotropes and intravenous fluid may be needed to correct hypotension resulting from a lack of sympathetic tone.<sup>20</sup>

The NHSBT service monitors adherence to rules for organ allocation, which are determined by the medical profession with the Department of Health and specialist advisory

**Box 4 | Questions to consider at retrieval of tissue**

Responses must be in the affirmative before tissue retrieval can legally occur.

Is the establishment where retrieval of tissue is to take place licensed by the Human Tissue Authority pursuant to the Human Tissue (Quality and Safety for Human Application) Regulations 2007?

If not, is there a third party agreement (drawn up in accordance with Directions 02/2007 issued by the Human Tissue Authority) between the establishment where tissue is to be retrieved and that establishment responsible for testing, processing, storage, and distribution of tissue?

If the material is to be retrieved during the course of a post mortem examination, do the premises where that post mortem examination is to be made hold a licence for the making of that examination?

Does the person retrieving the material have appropriate training and experience?

Has the person retrieving the material had sight of the consent to retrieval?

Has the person from whom the material is to be retrieved been identified properly to the person making the retrieval?

On examining the body from which material is to be retrieved, is there nothing which makes the person retrieving the material concerned that such material may pose a risk when transplanted into a recipient?

If no ante-mortem blood sample is available, and if there had been no transfusion in the 48 hours before death which might have resulted in 50% dilution of blood volume, has less than 24 hours elapsed since determination of fact of death?

If it is proposed to store the retrieved material for more than 48 hours (and if that material is not blood), do the premises where that storage is to occur have a licence for that storage?

Has the appropriate information been given to NHSBT?

groups. This system aims to treat patients equally, to ensure that organs are allocated fairly according to patients' needs, and to achieve the best possible match between donors and recipients. Such matching takes into account the age, size, and blood group of both the donor and recipient; for renal transplants, human leucocyte antigen (HLA) compatibility is also a consideration. The specialist nurse for organ donation can access a database that will identify the best matched recipient or the transplant unit to which the organ should be offered.<sup>21</sup>

Once organs have been allocated, retrieval teams from the transplant centre are dispatched to the referring hospital where the donor operation takes place. To ensure that the quality of transplantable organs is not compromised during retrieval, this operation is undertaken by an experienced surgical team. Typically such a team would comprise (as a minimum) a lead surgeon (consultant or a trainee in a higher surgical training programme), an assistant surgeon, a perfusionist, and a scrub nurse.<sup>22</sup>

Tissue retrieval is subject to specific regulations.<sup>23</sup> Box 4 lists questions that need to be considered by the person retrieving tissue, who may be a nurse, a pathologist, or other appropriately trained person.

Following this operation the incisions are closed and the body is transferred to an appropriate room for laying out and viewing by the family.

**Step 7: follow-up**

The family is offered follow-up by the specialist nurse either through a home visit or telephone call: typically a letter regarding the anonymised details of recipients is sent to family around two weeks after donation.

**What are the challenges?**

In 2008, in response to relatively poor rates of donation in the UK and as part of a radical overhaul, the Organ Donation Taskforce recommended that all parts of the NHS embrace

organ donation as a usual rather than an unusual event and make it a routine part of end of life management.<sup>2</sup> Junior doctors working on general wards may find it particularly challenging to implement this recommendation. Although large intensive care units invariably have well established (and practised) policies to manage potential donors, this may not be true for smaller units and general wards. Junior doctors may be uncertain about how to broach this issue with consultants and how to seek consent from patients and bereaved families. Consultant led ward rounds present an opportunity, but the issue is sufficiently important to be discussed with senior colleagues at any time. The quality of this discussion can be enhanced if the junior doctor has already conferred with the specialist nurse for organ donation about the suitability of a potential donor. This nurse is an excellent source of information about all aspects of donation and can be contacted via the hospital switchboard 24 hours a day.

Generally senior doctors or specialist nurses take the lead in seeking consent for donation from families (or patients) but trainee doctors should take the opportunity to learn by sitting in on these discussions. Factors that have been shown to increase consent rates for donation after brain stem death include the family's perception of the quality of care that their relative has received and their understanding of brain stem death.<sup>8</sup> Junior doctors can positively influence both of these aspects.

Junior doctors often lack suitable education and training about donation and transplantation. These topics are not prominent in the curriculums of most UK medical schools, and the current consensus statement on teaching medical ethics and law does not refer to organ donation.<sup>24</sup> It is also possible for junior doctors to complete training without being involved in the management of potential donors or being aware of donation requirements.<sup>2</sup> Increasing opportunities for them to work in intensive care units and emergency departments with significant experience in donation will help address some of these deficiencies. The organ donation taskforce has stated its strong support for the introduction of organ donation into the curriculums of medical and nursing schools.<sup>2</sup>

Balancing conflicting obligations in interpreting the best interests of the dying patient and potential organ recipients (so called dual advocacy) poses a difficult ethical and legal challenge.<sup>25</sup> In the UK, department of health guidance emphasises that if an individual has expressed a wish to be an organ donor then interventions to facilitate donation may be considered in that person's best interests, provided that they do not cause any harm or distress to that person.<sup>26</sup>

It is perhaps easy to understand why junior doctors, faced with these challenges against time constraints, may not engage in this process. We suggest that juniors keep in mind the life saving and transforming consequences of transplantation and actively seek advice from senior colleagues and specialist nurses when they are engaged in end of life management.

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## A PATIENT'S JOURNEY

# Duchenne muscular dystrophy

Stefan Spies,<sup>1</sup> Karen Schipper,<sup>1,2</sup> Frans Nollet,<sup>3</sup> Tineke A Abma<sup>2</sup>

Stefan is 27 and has Duchenne muscular disease; he lives in a residential home. Almost all his muscles have deteriorated; he can move his head slightly, has a lot of pain, needs artificial respiration 24 hours a day, and is almost totally dependent on others

I was diagnosed with Duchenne muscular dystrophy at the age of 3. My parents were always honest about my disease, but I didn't really care much about it. As the progression was slow, I gradually began to understand its impact, including the physical restrictions. I'm now severely disabled and have lived much longer than the doctors expected. The loss of opportunities open to me, combined with the fact that I'm still alive, evokes many mixed feelings.

### Moving to a residential home

I moved into a residential home when I was about 20 years old. This wasn't easy. I had to get used to being helped by a succession of professionals. Until then, my mother had been my only carer. The move was also difficult for my mum. She had to trust new, unknown, professional

### HELPING HANDS ALONG THE WAY

People who support me by asking how they can help me and by asking if I need or want help. Sometimes people have good intentions, but are clumsy in their actions

The support of my parents, family, and friends, and of my religion

My willpower and persistence and my ability to see the bright side of things again and again

Taking responsibility for my own behaviour and my relations with others. We have to care for each other

My wish to motivate other people and my drive to teach them something about life. This is the power of giving. By doing this, I can make sense of my illness

caregivers. It was hard for me, too, since I love my mum and I want her to be happy.

### Constant adjustments

I have to adjust repeatedly to changing reality. I used to play wheelchair hockey, but I'm not able to do that any more. I cannot go out without help. My ideas and preferences have also changed. Initially, for example, I resisted when the doctors said I needed artificial respiration during the night. I accepted this for night time, but didn't

<sup>1</sup>Reinaerde, Postbox 3364, 3502 GJ Utrecht, Netherlands

<sup>2</sup>VU Medical Centre, EMGO Institute, Department of Medical Humanities, Postbox 7057, 1007 MB Amsterdam, Netherlands

<sup>3</sup>Department of Rehabilitation, Academic Medical Centre, Postbox 22660, 1100 DD Amsterdam, Netherlands

Correspondence to: K Schipper k.schipper@vumc.nl, Frans Nollet f.nollet@amc.nl

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This is one of a series of occasional articles by patients about their experiences that offer lessons to doctors. The *BMJ* welcomes contributions to the series. Please contact Peter Lapsley (plapsley@bmj.com) for guidance.

### A DOCTOR'S PERSPECTIVE

Duchenne muscular dystrophy is a progressive muscle disease affecting boys. The muscles degenerate as a result of defective dystrophin caused by mutations on the X chromosome. The typical course of the disease is delayed motor development that increases. Diagnosis is usually made at around preschool age. As muscle weakness progresses, walking becomes more difficult. Around the age of 12, the boys become wheelchair dependent. Later on, arm and hand functions decrease, and problems with swallowing and speech arise. Respiratory insufficiency in the late teens causes death unless mechanical ventilation is applied. Mechanical ventilation has become a routine part of treatment over recent decades.

Boys with Duchenne muscular dystrophy are confronted with many medical issues as they grow up, such as possible cardiac involvement, the need for scoliosis surgery, side effects of corticosteroids like osteoporosis, and growth retardation. They become increasingly dependent on orthoses and wheelchairs, and on personal help. In childhood the parents are the most important caregivers.

Stefan, a 27 year old man with the disease, has many worries and fears—about his end of life, about the possibility that his ventilator may become defective, and many other issues. He finds it difficult to adjust to changes. Not wanting to be a burden is a common attitude among young people who have always been dependent on others. This hinders the sharing of concerns with caregivers and intimates. They already have to help so much, so Stefan thinks he should not bother them with his troubles. Knowing that feelings are not easily voiced, explicit attention should be given so that people like Stefan have a trusted person they can talk with, preferably someone they are not dependent on.

With prolonged life expectancy, children with Duchenne muscular dystrophy nowadays make the transition into adulthood and often move to specific housing where help is given by professional caregivers. Stefan makes clear that this is not an easy change. Although help is provided, professional caregivers have different routines and opinions, and are not as trusted as the mother. This complaint of differences in behaviour between caregivers is common. Caregivers should act more consistently to accord with the individual's needs. This should be taught, learnt, and discussed openly with the patient.

Stefan has said that he has a lot of pain; whether this is dealt with adequately is not clear. The problem of pain in Duchenne muscular dystrophy is underestimated by health professionals, and it is conceivable that Stefan will not raise the issue easily.

The risk of treating children and young adults with Duchenne muscular dystrophy is that, with so many medical and rehabilitation issues to deal with in an ongoing chain of events, the person involved may easily be overlooked. Measures are applied by many health professionals, all intended to help (see box). Deciding what needs to be done should be based on guidelines. As the child changes into a young adult with his own opinions, needs, and concerns, health professionals need to change their attitude accordingly, from deciding for the child to listening to the patient and responding to his needs.

Frans Nollet f.nollet@amc.nl

### WHAT CAN MEDICAL PROFESSIONALS DO?

- Give patients and their relatives enough time to adjust and to get used to the idea of impending changes
- Give emotional and mental support. Doctors are often too busy with the medical aspects of the disease; they should pay more attention to the whole patient and his emotional wellbeing. Patients need somebody who really listens to them. Living with Duchenne means continuous adaptation and continuous regression—because of this, continuous emotional support is needed
- Professionals try to find solutions for problems. The solution to a practical problem, like a new alarm system, is not the same as the solution for the underlying emotional difficulties. These emotions cannot be solved with technical or practical solutions, but they also need attention
- Living with Duchenne evokes many existential questions. Those questions can't be solved alone or with the help of daily caregivers. Psychosocial support is needed to deal with clients' questions and emotions
- Practical support should be attuned to individual needs and wishes
- The whole family should have support, since Duchenne is difficult for all family members
- Some patients and family members wrestle with feelings of responsibility. Professionals should be aware of this and should support patients and family members who express responsibilities felt for others

want a cannula 24 hours a day. With a cannula I would be totally dependent on others. But when eventually I actually needed respiration 24 hours a day, I agreed. It scared me and at that moment I wished I was dead. It was an enormous step, but my feelings changed over the time. Still, it is hard, every time a new problem occurs.

I need some time to get adjusted to the changes, and medical specialists sometimes go too fast. The operation of the cannula was planned three weeks after the decision, which gave me some time to adjust—but then I was asked to undergo it earlier. Medically that was probably a good decision, but for me it was too sudden. I was upset and thought about cancelling the operation. Afterwards I was happy with the change, but not knowing what's going to happen is scary.

### Fear and dependency

For me, the fear and dependency is the worst. I am totally dependent on others and can't do anything at all. Were something to go wrong with the cannula, I would be able to survive for only three minutes without artificial respiration, then I would die.

An automatic alarm is set to go off in case of an emergency, or I can use the alarm system myself. It frightens me—does it work? Will the caregivers be in time? Sometimes, the plug for the alarm bell comes out for a little during the night. I can't use it if this happens, since I'm not able to put it back, so I have to scream. I'm afraid there may come a time when they don't hear me.

Artificial respiration restricts me. I can't go out alone, I need somebody to help me in an emergency and to drain off the mucus in my trachea.

Some people with artificial respiration go outside without other people. They ask a passer-by in case of an emergency or if they need help. I don't do that since I don't want to be troublesome and I don't want to take risks. I want somebody to go with me who knows how to respond properly if necessary.

I do not want to be troublesome in my home. I try to remember the answers to my questions and ask for help if I have more questions. Sometimes I lie uncomfortable in bed for hours, because I don't want to call for help. This is not satisfactory either.

### Religion

My religion really helps me. I'm not angry because of my disease but I'm disappointed if things go wrong. I realise that the Lord will support me always. Maybe not immediately, but later on. I realise that He was with me in these difficult moments.

Of course, I wish I could recover, but that does not appear to be a possibility. People in church pray for me and for my recovery. I think it's really sweet, but for me my disease also has a meaning. It's not senseless, since I'm now able to show people how privileged they are. My body tells them that being healthy isn't something to be taken for granted.

I think we should realise that we are all vulnerable. People have to know that they could be disabled the next day without warning. My disease can motivate people to create a good life, to make them realise that life isn't always easy.

### Together but lonely

I have many friends, and they and my family are important to me. Other people sometimes have problems with handling my disability. Because of this, they avoid talking with me and find it difficult to gauge my mood and situation.

Often I don't want to talk about my disease or express my feelings. I keep my fears, doubts, and questions to myself as I don't want to make people feel sad. I want it to be cosy. Sometimes this sense of responsibility is tiresome. Then I can't let it go, which leads to lying awake, thinking about it, at night, and makes me feel sad and angry.

### Relations and sexuality

I would like to have a wife, but then I also would long for children, and having children is impossible. I wouldn't be able to handle situations like being unable to hold my child or for us to play together.

I think my wife shouldn't have to be responsible for the children and me, and to know that she would end her life alone, without me. That makes me sad. And because of this, I don't want to marry. But I do have some sexual needs and I don't know what my religion allows. If I watch a movie, I feel guilty and sad afterwards. My religion supports me, but during these moments it's difficult. Also, it's hard seeing friends and family getting married and having children, and realising that I will die without such things.

### Empty days and dreams for the future

My days are long and I'm at home most of the time. It's boring. I watch some DVDs and I surf the internet. I read

in the Bible and think about my life and what's going on in the world. I would like to go to a day activity centre again. This is only possible with a wheelchair taxi and in company with a skilled person. Unfortunately that's expensive. We've had some activities in the living room of our home, which was great. At the moment, the ground floor is being rebuilt to create a real day activity centre. I'm looking forward to visiting it and meeting other people.

### The end of my life

I try to enjoy life as much as possible, but for the last few years I have been thinking more and more about the fact that I'll die early. It makes me sad. I have already lived longer than everybody expected when I was born.

I'm scared about the future and about dying, so I try to focus on the nice things in the future. And even if it's hard, you have to go on. Sometimes I wish it could be all over, but at such times I can always find a bright spot again.

**Contributors:** Because SS is incapacitated, he told his story to KS during an interview. KS recorded SS's experiences and used his words to draft the article in the first person. SS read the article and adjustments were made by KS until SS recognised it as his own story. TAA helped KS with the writing process and commented on and edited the text. KS translated the article into English, with TAA's help. FN supplied the doctor's perspective. KS is guarantor.

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## ANSWERS TO ENDGAMES, p 1109. For long answers go to the Education channel on [bmj.com](http://bmj.com)

### STATISTICAL QUESTION

#### Skewed distributions II

Answers *a*, *b*, and *c* are true, whereas *d* is false.

### ON EXAMINATION QUIZ

#### Thyroid dysfunction

Answer D is correct.

### PICTURE QUIZ Doc, my knees give way!



Fig 1 | Lateral view radiograph of the patient's left knee showing suprapatellar swelling, quadriceps disruption, and suprapatellar calcification



Fig 2 | Lateral view radiograph of the patient's right knee showing suprapatellar swelling, quadriceps disruption, and suprapatellar calcification

- 1 The two lateral view radiographs show soft tissue swelling, disruption at the quadriceps tendon, and suprapatellar calcific density. The patellae are low lying.
- 2 The most likely diagnosis is bilateral rupture of the quadriceps tendons. Other possible diagnoses that need to be considered are patellar tendon rupture and injuries to the menisci or anterior cruciate ligament.
- 3 The clinical findings of a palpable gap proximal to the patella and knee extensor lag are classic signs of bilateral rupture of the quadriceps tendons, as are absent patellar reflexes.
- 4 The patient should undergo surgery to repair the ruptured tendons and be kept immobilised with his knees in extension for six to eight weeks, then should undergo strict physiotherapy to regain strength and range of motion.