

A PATIENT'S JOURNEY

Visual agnosia

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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 (lapsley@bmj.com) for guidance.

This patient was left with visual agnosia after developing herpes simplex encephalitis at a young age. She describes her strategies for coping with this visual disability

When I was 11 months old I developed herpes simplex encephalitis after coming into contact with a cold sore, which damaged the areas of the brain responsible for processing vision. I was left with visual agnosia and had severe difficulties in recognising people by their faces. I cannot remember the details of the earliest events surrounding my illness, but I have talked about them a lot with my family and this is my story.

During the illness I lost my sight as well as my ability to sit and walk. It took me about six months to relearn to walk unaided. During this time my vision gradually improved, but I had to rely on my other senses to interact with the environment. I would sniff, touch, and taste things around me, including the floor, furniture, and people's clothing, which some found unnerving.

My speech development was good. I found changes in routine upsetting and puzzling. Mum had the support of medical and educational professionals and on their advice I was taught at home by an educational service (Portage) for preschool children with additional support needs and their families, and I had a support assistant at play school. Around this time mum noticed I was making mistakes in recognising family members. Once I was seen talking to my brother's friend, mistaking him for my brother (who looks completely different).

It wasn't just a problem with recognising family members, or people in general—mum had painted a Postman Pat mural on my wall and despite being familiar with his character through books and DVDs I just could not recognise him. However, it was when mum realised that I couldn't even recognise her that things came to a head. This happened on a Portage course for parents. As the parents entered the crèche, the children got up and went to their mums and dads. I got up too, studied the adults and then returned to my carer rather than going to my mum. Although mum had been standing close to me, it wasn't until she spoke that I got up and took her hand. Mum talked about this to a teacher for visually impaired people at my nursery. He said he thought I might have a facial agnosia. This was discussed with the child neurologist at my next appointment, and subsequently with a neuropsychologist.

My lack of recognition of family members caused all sorts of difficulties. Mum will never forget the time I was on a child's amusement ride in the supermarket while she was close by paying for her shopping. When the ride had finished she heard me calling for her and then saw me being lifted out by a woman. This stranger was reluctant to hand me back to my mum because she didn't see the flash of recognition she was expecting from me.

I also had problems with object recognition. No one could understand how I managed to find small objects dropped on the floor but wasn't able to recognise pictures of everyday objects. When given a verbal clue I would always get the object right. I couldn't visually distinguish between animals such as a cat and dog or a cow and sheep. This problem only improved after many visits to farms and zoos.

When I first started school my eye contact was poor and despite constant encouragement from my family, teachers, and carers to lift my head I found it hard to look at people. My confidence was lowered further by the repercussions when I misread facial expressions.

Some aspects of my condition have improved but not the facial agnosia and I continue to find the recognition and identification of faces difficult. Recognition is even harder when people are out of context or a particular feature such as hair colour has changed. School was on occasions frustrating, not least because everyone wore the same uniform. Finding friends both in and out of the classroom could be difficult, so I had to rely on my other senses. Things became easier when I moved into sixth form, as students could wear casual clothing. Teachers found my disability difficult to understand. Some believed it didn't exist and others didn't understand that the problem arose from my brain not my eyes. Unbelievably, at a parents' evening a teacher asked mum whether all her children were blind, to which (after taking a deep breath) she replied "On my last count, none of my children is blind."

A DOCTOR'S PERSPECTIVE

Visual agnosia is a specific failure of visual recognition of objects not explained by elementary visual deficits. The agnosia can be category specific—for example, prosopagnosia is a specific agnosia for faces. Pure visual agnosia is a rare condition, particularly in young people, but visual agnosias can also occur as part of neurodegenerative conditions.

This patient's visual agnosia was secondary to early acquired herpes simplex encephalitis, which caused bilateral damage to the occipitotemporal regions. Those brain areas specifically involved in processing faces were affected, and although the patient made a good recovery overall, face recognition remains problematic on a day to day basis. Her story is remarkable on several levels. One of the most striking aspects is the "invisible" nature of the condition. It is not surprising that people may be confused by the fact that someone can see well enough to go horse riding but cannot distinguish between family members by their faces. What people don't understand they often refuse to believe and this has clearly led to some awkward situations. This patient's drive to study the condition in detail and to explain it to others is both understandable and commendable. Her ability to perceive the world differently can also be viewed as a strength and has led to some original artwork.

Doctors can help those with visual agnosia by being alert to the possibility of disorders of higher visual function, facilitating appropriate neuropsychological assessments, and providing explanations and accessible strategies for families and schools. To achieve all this, however, requires awareness of such conditions.

Most remarkable of all are the sheer courage and determination shown by this patient and her family to help overcome her difficulties, enabling her to succeed both in and out of school and in holding down a job. I have learned much from sharing this part of her journey and am proud to know her.

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Previous articles in this series

► Lessons from patients' journeys
(*BMJ* 2013;346:f1988)

► Klinefelter's syndrome—a diagnosis mislaid for 46 years
(*BMJ* 2012;345:e6938)

► Kallmann syndrome
(*BMJ* 2012;345:e6971)

► Non-coeliac gluten sensitivity
(*BMJ* 2012;345:e7982)
► Restless legs syndrome
(*BMJ* 2012;345:e7592)

Getting teachers to complete the simplest of tasks, such as using a black pen on the whiteboard and enlarging worksheets, was a test all of its own and made life in the classroom problematic. Despite these challenges I was successful at GCSE level, with 10 good grades. Subsequently I studied English language, art, and psychology at A level. As part of my psychology course I studied face perception and its disorders and was asked to give a series of talks to the other psychology classes. For my art A level I brought together elements of Cubism and still life and incorporated texture to give my work a more original and interesting direction, reflecting the importance of touch in helping me to understand my surroundings in my early life.

Over time I have built up my own strategies for recognising my family and friends and those I am in daily contact with like my work colleagues; for example, by remembering their style of walk, hairstyle, and hair colour. I have also become good at recognising voices and scents, including the perfume worn by particular people. Recently I have become more sensitive to sounds such as those made by jewellery and keys. I am able to find mum in crowded places by the sound of her bangles and dad by the sound of his car keys. This has increased my confidence in public places. I remember when I was around seven years old being perturbed at meeting identical twins who were also identically dressed. Although neither I nor anybody else could distinguish the twins by sight, for once my personal strategies for distinguishing between people by voice, scent, etc, failed me. I was left so scarred by this experience that months later I avoided a close family friend "because she might have a twin."

A few years ago I was approached by my neurologist, Professor Eyre. She asked if I would take part in a research study of face training, with the aim of relearning the mathematics of face recognition and using func-

USEFUL RESOURCES FOR PATIENTS AND CLINICIANS

Farah M. *Visual agnosia*. 2nd ed. MIT Press, 2004—A classic text on disorders of visual recognition

National Portage Association (www.portage.org.uk/)—A home-visiting educational service for preschool children with additional support needs and their families

National Institute of Neurological Disorders and Stroke (www.ninds.nih.gov/disorders/prosopagnosia/Prosopagnosia.htm)—Patient information on prosopagnosia, or "face-blindness"

Faceblind.org (www.faceblind.org/)—Information from researchers and opportunities for those affected to take part in research

tional magnetic resonance imaging scans to look for any changes in the brain. When babies and children study faces intently their brains are learning to calculate the distance between eyes, nose, and mouth and the width and length of faces, which are the basics of expertise in recognising faces. The training programme required me to categorise faces based on differences in these distances. Involvement in the study has improved my eye contact and confidence. My training scores have also improved but I still have trouble recognising faces. Despite this I am becoming more confident in my day to day recognition of friends and family using all available means.

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10-MINUTE CONSULTATION

Vasectomy

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This is part of a series of occasional articles on common problems in primary care. The *BMJ* welcomes contributions from GPs.

A 40 year old man attends your clinic with his 37 year old wife to discuss long term contraceptive options. They inform you that they have three children and have completed their family. The wife has been taking oral contraception for the past five years but does not want to continue with this. They have both considered other contraceptive options and have decided on vasectomy.

What you should cover

Patient selection and counselling is crucial. To determine if vasectomy is suitable for them, consider the following points.

- Ideally both partners should be present.
- Establish the patient's age, relationship status or stability, and number of children. Young, single men with no children are more likely to regret their decision and request a reversal later in life.

- Discuss other contraceptive methods. Female sterilisation (tubal ligation) can be done hysteroscopically under local anaesthesia; or laparoscopically or through a mini-laparotomy, under general anaesthesia. Consequently, the morbidity of the procedure is higher than for vasectomy, and some studies suggest that the reported lifetime failure rate is higher, at 1 in 200.
- Emphasise the need to use alternative contraception after vasectomy until semen analysis confirms the absence of sperm (usually three months).
- Ask about any current systemic or sexually transmitted infection, coagulation or other blood disorders, and chronic testicular pain, as these will influence the timing of and preparation for surgery. A small number of patients will develop chronic testicular pain after this procedure, which can interfere with their quality of life. This typically occurs in 1-2% of men.

Complications

Vasectomy has an associated low complication rate; patients should be informed about the following complications.

Common

- Scrotal bruising, haematoma
- Chronic testis pain affecting quality of life (1-2%) and sperm granuloma

Occasional

- Bleeding requiring further surgery

Rare

- Infection of testis or epididymis requiring antibiotics
- Early recanalisation resulting in persistence of motile sperm in the ejaculate for which repeat vasectomy is indicated (0.2-5.3%)
- A risk of about 1 in 2000 of pregnancy resulting from late recanalisation after previous clearance

FURTHER READING

For patients

- Patient.co.uk (www.patient.co.uk/health/Vasectomy.htm)
- NHS Choices (www.nhs.uk/conditions/vasectomy/pages/introduction2.aspx)
- British Association of Urological Surgeons (www.baus.org.uk/Resources/BAUS/Documents/PDF%20Documents/Patient%20information/Vasectomy.pdf)

For healthcare professionals

- European Association of Urology Guidelines on Vasectomy (www.europeanurology.com/article/S0302-2838%2811%2901101-8/fulltext)
- American Urological Association Guidelines on Vasectomy (www.auanet.org/content/media/vasectomy.pdf)

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Previous articles in this series

- Assessment and management of renal colic (*BMJ* 2013;346:f985)
- Dry eye (*BMJ* 2012;345:e7533)
- Minor incised traumatic laceration (*BMJ* 2012;345:e6824)
- Adult trigger finger (*BMJ* 2012;345:e5743)
- Myalgia while taking statins (*BMJ* 2012;345:e5348)

- Examine the testis and both vasa deferentia for any abnormality that may interfere with the procedure, such as a large hydrocele, inguinoscrotal hernia, or testis tumour.
- Emphasise the need for post-vasectomy semen analysis three months after vasectomy. Some sperm may still be present in the semen for a few months, and so the patient should continue other methods of contraception until the semen analyses show azoospermia.
- Inform the patient that 100% guarantee cannot be given of permanent sterility as there is a rare chance of early failure (0.2-5.3%) and late recanalisation (0.03-1.2%).
- Explain the operation and complications (box).
- Provide the couple with leaflets or booklets or other printed material outlining the information covered in the counselling session.
- If there is any doubt, ask the couple to return for a further consultation once they have had time to make their decision.

How it works

Vasectomy is an effective, reliable, and permanent form of male sterilisation with no serious long term side effects. It is usually done under local anaesthesia and very occasionally under general anaesthesia. In the conventional incisional technique one or two small incisions are made with a scalpel on the scrotum and both vasa deferentia are exposed. The no-scalpel method involves puncturing the skin with a sharp haemostat. A small segment of vas is removed from both sides and the ends are ligated with sutures. Soft tissue is interposed between the two ends of vas to prevent recanalisation.

FREQUENTLY ASKED QUESTIONS

Q: How effective is vasectomy?

A: Vasectomy is a safe and reliable procedure. However, 1 in 2000 men can become fertile again due to rejoining of the two ends of the vas, so vasectomy is not 100% effective.

Q: Can I stop using contraception immediately after the procedure?

A: No. Sperm can still be present for a few months and so a semen analysis is done three months after vasectomy. If it is clear of sperm you can stop using contraception.

Q: Will it affect my sex drive?

A: No. The sex hormones will still be produced by the testes so libido and erectile function are unaffected.

Q: What will happen to the sperm?

A: Sperm will still be produced by the testis but will get dissolved in the body.

Q: What if I change my mind after the operation?

A: You should regard vasectomy as a permanent procedure. The success rate of vasectomy reversal is only 30-60%, with a lower success rate in men who have a later reversal.

Q: Does the operation hurt?

A: It is done under local anaesthesia. You may experience some discomfort during the injection and the procedure but not severe pain.

Q: How soon after the operation can I have sexual intercourse?

A: You can resume sexual activity as soon as it is comfortable. You have to use other methods of contraception until the semen analysis shows no sperm in the specimen.

Q: Is there any risk of prostate or testis cancer after vasectomy?

A: No. Several studies have shown no relation between vasectomy and cancer.

Aftercare

After the procedure there is usually some discomfort and bruising for a few days, which can be improved by wearing tight fitting underwear. Patients are advised not to work on the day after the operation and that a return to normal activity is usual within a week. Three months after vasectomy a semen analysis is required. If no spermatozoa are detected in the ejaculate, give the patient clearance to stop using other methods of contraception. If motile spermatozoa persist at the six month follow-up, a repeat vasectomy is advisable. Other contraceptive methods should be continued until clearance is given.

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