

## Babington's aphasia— overcoming left hemisphere trauma

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*J R Soc Med* 2003;96:23–27

Anthony Babington was 24 and a company commander in the 1st Dorset Regiment when he was wounded by shrapnel in Holland early in the morning of 2 November 1944, in the aftermath of the Arnhem campaign. His story is in his own writings<sup>1,2</sup> and in his army medical records<sup>3</sup>, both quite exceptional in detail and perception.

### CASE HISTORY

He was felled by the shell burst and briefly unconscious, and had sustained a left parietal penetrating injury which made him mute and hemiplegic. He was evacuated by stretcher, ambulance and plane. 'During the flight I felt something stiff and cold on the right side of my chest. I puzzled about it for a while before it dawned on me that it was my right arm, heavy, insensate and quite paralysed . . . At one point I thought the end had come . . .'<sup>2</sup> On the evening of the same day he reached the Military Hospital for Head Injuries at St Hugh's College, Oxford.

X-rays showed 'a large MFB [metal foreign body] deep in the left parietal lobe just above the ventricular roof but may impinge on it.' On 3 November Major C A Calvert<sup>4</sup> operated on a semiconscious, globally aphasic man who had a right hemiplegia, hemianaesthesia, and hemianopia (Figure 1):

' . . . wound stank horribly . . . contamination with hairs and dura down brain track—marked oedema . . . toothpaste brain and clot . . . bone chips indrawn about 2 cm, metallic body about 4 cm from cortex, inner end about 6 cm . . . debrided . . . many toothpaste pockets extending laterally from track . . . area of cortex destruction about 4 cm×4 cm . . . gush of CSF . . . ventricle into which MFB had protruded . . . 5000 U penicillin . . . dura left open . . . drain . . .'<sup>3</sup>

After the operation Anthony Babington recalled<sup>1,2</sup> gesticulating with his left hand to indicate that he needed treatment for head pain, something to drink, or a urine bottle. When asked to write his home address with his left hand 'I knew exactly what I intended to write but I was

dismayed to find myself drawing a series of short and very crooked lines . . . when shown a newspaper I could see letters but not understand them . . . might have been Chinese.

'I woke one morning feeling slightly better. The night sister said "You are over the worst and out of danger . . . The doctors think you might get back a little movement in your right side, in the leg at any rate. But I am afraid they think it unlikely that you will ever be able to speak again".'

This forecast also ended a close personal relationship.<sup>2</sup>

Like Dr Johnson after his stroke<sup>5</sup>, he was comforted by his ability to say the Lord's Prayer silently.

In the medical records<sup>3</sup> Captain G Northcroft wrote on the fourth postoperative day that there was now no hemianopia and that he could indicate yes and no by head movements. On 11 November he could just utter 'well', and on the 14th there were a few single words, and 'speech therapy' was started by a sympathetic lady who had trained as a singing teacher and by a Norwegian fellow patient<sup>1,2</sup>.

### Early progress

He began to try to read a book: ' . . . laborious . . . words slowly . . . wrong lines . . . Writing even more discouraging . . . left hand awkward . . . entirely lost the art of spelling . . . even ridiculously simple words. I did not seem capable of visualising them on paper'<sup>1</sup>. Three months later his speaking, reading and left-handed writing had improved enough to make feasible his intention to take up law studies, according to psychometric testing by Major W R Reynell<sup>3</sup>. He was discharged from hospital in May 1945, with the advice to take up market gardening. But he went to the Middle Temple later in 1945, and in 1948 he qualified as a barrister, having had the use of an amanuensis for his written examinations<sup>1</sup>.

### The next 57 years

Ever since, his speech has continued to be slow, with a slight stammer, and persistent inability to utter some words which he has managed to cover by circumlocution; there are no wrong words. His reading also has remained slow, both silent reading and reading aloud, but adequate for public performance. He achieves a slow sinistral script which looks like his old dextral writing but with persistent mis-spelling, even of common words (Figure 2). He has been rendered moderately dyscalculic by the injury, but considers his powers of reasoning, and his memory for abstract data, for places and faces, as good as before. He never experienced difficulty in understanding spoken speech. He had a successful career as a barrister, at first in chambers, then

70/1275 28.000 9/43 A. & E.W.Ltd Gp. 698  
40/752 42.000 2/44

Army Form W 3118

## Operation Card for Head Injuries

(For distribution to Casualty Clearing Stations, Stationary and Base Hospitals.)

(Field Card No. W. 3118B should be filled in before operation)

Name Capt. Ballington Rank Capt.

Unit \_\_\_\_\_

Date of operation 3rd Nov. 44 09.30 hrs Time (in hours) between injury and operation \_\_\_\_\_

Situation of wounds (enter on diagrams) Left parietal 8.5 cms above ear and along meatus

X-ray findings. Fracture? about 2 x 2 cms F.B.? Yes Site of F.B.? L Parietal lobe above 3.5 cms deep to Rolandic

(Mark X-ray findings on diagrams)

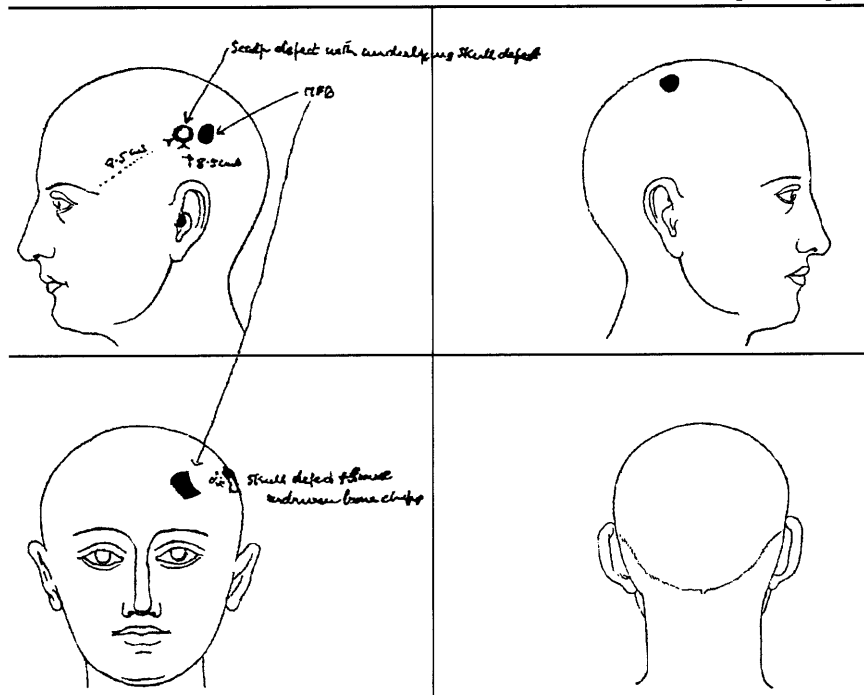


Figure 1 C A Calvert's operation diagram of 3 November with measurements of site of lesions from X-rays

for 8 years as metropolitan magistrate, and lastly as circuit judge until beyond the usual retiring age<sup>2</sup>.

He has also written ten books, three of which were republished. Two are autobiographical<sup>1,2</sup>, the others are on legal and military history. His writings were influential in changing the public attitude to the execution of soldiers in the First World War for desertion and for cowardice, and to 'shell shock'.

He had bad headaches in the first post-traumatic year, and intermittently later, related to low-grade skull infections until the tantalum skull implant was removed by Mr C Connolly in 1975. In the early years he had a few partial (right hemisensory) seizures, treated with phenobarbitone; he stopped taking this because of retardation, and the attacks have remained in abeyance. The right hemisensory loss has persisted, as has near-total paralysis of the right

upper limb. The initial right lower limb paralysis improved sufficiently to allow pleasurable walking of up to five miles, but has worsened in his 70s, and he is now limited to a slow few hundred yards with a walking-stick. He also weathered long spells of inpatient treatment, with a lobectomy in 1951, for pulmonary tuberculosis, and later acute surgery for a duodenal ulcer, and aortic valve surgery. Despite his disabilities he retains his *joie de vivre* at the age of 82.

### COMMENT

#### Site of lesion and clinical features

Speedy and expert neurosurgery, as well as the newly available penicillin, were lifesaving. But he was left with a deficit of parietal cortex ('4 cm across') extending deeply through the white matter to the lateral ventricle.

(2) Mental I find reading troublesome I can only read slowly and frequently misread sentences. I have a difficulty at first with spelling when I write. This is now much improved but I still suffer from word blindness. I have a tendency to get depressed about my condition. This is now

1946

He concludes that there is nothing wrong with the foot and the splint is ~~useful~~ <sup>useless</sup>. He arranged for me to see ~~one~~ <sup>one</sup> of the consultants and the chief physiotherapist

I saw them last

1994

Figure 2 Dysgraphia in 1946 and 1994: spelling problems and elisions

There must also have been damage to the left (frontal) motor cortex, or to its connections, to cause the right hemiplegia.

The initial aphasia was termed 'global'<sup>3</sup>, but he did not seem to lose understanding, even at his worst. Much of this, like the transient right hemianopia, must have been due to cerebral oedema. The penetrating shrapnel and bone chips impinged just in front of, and above, the temporal gyri described in 1874 by Wernicke<sup>6</sup> as the site of sensory (i.e. receptive) aphasia, and a little further from Broca's more anterior low frontal site of motor or expressive aphasia (Figure 3). Both Broca and Wernicke made their discoveries from cases of cerebrovascular disease; Broca in particular was severely criticized in 1906 by Pierre Marie<sup>7</sup> for anatomical oversimplification, as Broca's extant post-mortem specimen had shown more extensive lesions than just in the left second and third frontal convolutions.

Accordingly aphasiologists preferred the 'crisper' trauma material from the wars of the last two centuries—Larrey<sup>8</sup> from the Napoleonic wars, Henry Head<sup>9</sup>, Pierre Marie<sup>10</sup> and Kurt Goldstein<sup>11</sup> from the First World War, and A R Luria<sup>12</sup> and Ritchie Russell<sup>13</sup> from the Second World War. The studies of the last dealt with the 1100 head injuries treated at St Hugh's College, Oxford<sup>3</sup> and the other British neuro-surgical units.

While confirming the great importance of Broca's frontal and Wernicke's temporoparietal speech areas, both the Russian<sup>12</sup> and the British<sup>13</sup> compositions agreed on a wider left hemisphere distribution of lesions causing aphasia, but clustering around Broca's and Wernicke's areas, with, conversely, an absence of aphasia after more distant left hemisphere lesions (Figure 4). Right hemisphere lesions were exceptional in causing aphasia, even in left-handers. Freda



Figure 3 Centre of lesion in supramarginal gyrus at intersection of lines from diagram (see Figure 1).

C=Central sulcus; B=Broca's area; W=Wernicke's area: superimposed on sagittal MRI scan of normal brain

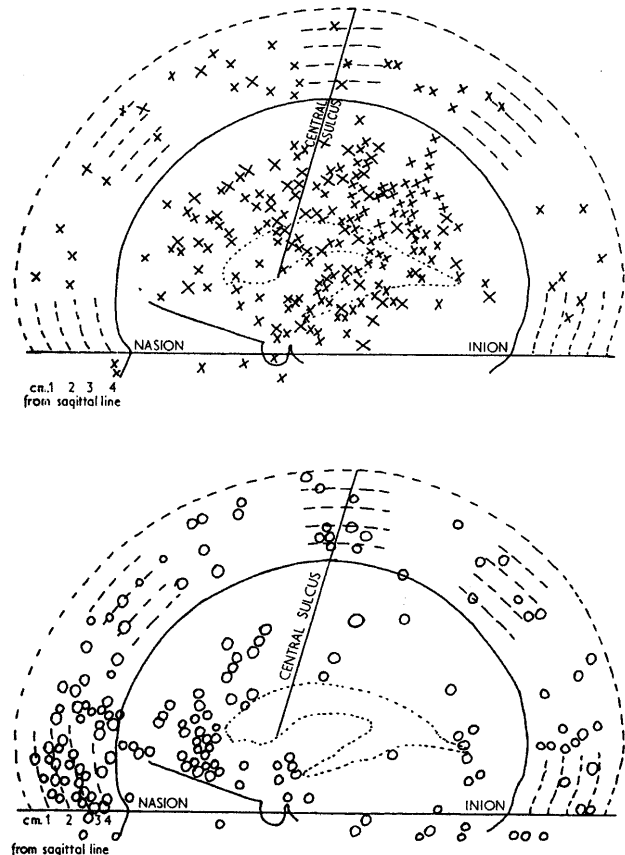


Figure 4 Composite diagrams of penetrating left hemisphere lesions causing aphasia (x), and not causing aphasia (o) (from Ref 13)

Newcombe<sup>14</sup> conducted a more intensive late assessment in 153 selected patients from the above 1100 military head injuries, noting in particular professional achievers. Anthony Babington is not discernible in her<sup>14</sup> or in Russell's<sup>13</sup> accounts.

## Recovery

Apart from the spontaneous resolution of brain oedema after C A Calvert's operation, leading to the improvement recorded in the first few weeks, his later recovery may be attributed in part to his determination and vocation. There is also the theoretical concept, in a young man, of reserve brain potential and 'plasticity'<sup>15</sup>, linked perhaps to the degree of left hemisphere dominance and right hemisphere speech functions<sup>16</sup>.

The notion of left hemisphere dominance goes back to the observations of Dax and Broca in the 19th century, and has an anatomical basis in specially developed areas of cortex on the left side<sup>17</sup>. While most left-handers, like right-handers, have their speech 'centres' on the left side of the brain, as judged by Wada's intracarotid sodium amytal injection testing<sup>18</sup>, they may still recover better than right-handers after left hemisphere lesions with aphasia<sup>12,19</sup>. One needs to consider degrees of left-handedness or of ambidexterity, and any family history of left-handedness, or left eye or ear dominance<sup>16</sup>. While dextrad for everything else, Anthony Babington was a left-handed bowler at cricket and a left-footed footballer as a boy.

Beside the overall better outlook in traumatic than in stroke aphasia<sup>20</sup>, Babington's high initial intelligence was probably also a good prognostic factor<sup>21</sup>, as opposed to the adverse prognostic factors of the immediate severity and 'globality' of the aphasia<sup>10</sup> and of the concomitant hemiplegia.

Did the so-called speech therapy help his recovery? While there has been continuing interest in communication skills after severe brain traumas<sup>22</sup>, there has been no critical study of the value of speech therapy in aphasia<sup>23</sup>. The bulk of aphasia cases in peace-time are due to strokes, with inevitably great variability and a worse prognosis than in the mostly younger sufferers of traumatic aphasia—and, indeed, amateurs may do as well as trained speech therapists in stroke aphasia<sup>24</sup>.

Modern investigations such as positron emission tomography (PET) scanning<sup>25</sup> and functional magnetic resonance imaging (fMRI)<sup>26</sup> which can demonstrate changes in regional cerebral blood flow during various language tasks have greatly advanced our ideas of the anatomy and physiology of speech, beyond what had been derived from the deficits resulting from brain lesions: it has unfortunately not been possible to study Anthony Babington's communication skills in this way.

That he achieved so much in his two vocations as a lawyer and as a writer may serve as a warning to dismal prognosticians, and as a tribute to his personality.

*Acknowledgments* I am most grateful to His Honour Anthony Babington for permission to write about him, and for his help; also to the late Dr Freda Newcombe for giving me access to the medical records at Oxford. I am obliged to David Jellinek and Charles Romanowski, of the neurosciences department, Sheffield, for demonstrating the neuroanatomy of the lesion by MRI superimposition.

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## Visual field compression by a non-secreting pituitary tumour during pregnancy

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The pituitary gland enlarges physiologically in pregnancy, mainly because of lactotroph hyperplasia<sup>1</sup>. Pre-existing pituitary tumours can likewise increase in size<sup>2</sup>, but compressive effects may be due to displacement rather than enlargement.

### CASE HISTORY

A woman of 29 was referred to the endocrine clinic for investigation and management of oligomenorrhoea and infertility. Adrenal function was normal, she was euthyroid, and visual fields were normal to confrontation. Investigations suggested anovulatory cycles in the presence of adequate oestrogens (serum prolactin 430 mU/L [normal range 150-800], testosterone 1.4 nmol/L [0.3-2.5], sex hormone binding globulin 130 nmol/L [30-90], oestradiol 300 pmol/L; a menstrual bleed followed administration of medroxyprogesterone acetate 10 mg daily for 5 days). She was treated with clomiphene 50 mg on days 5-10 of the cycle and became pregnant that month.

At 18 weeks' gestation she noticed abnormal vision in the left eye. On examination acuity was reduced on the left (6/12 compared with 6/5), with a central scotoma, loss of vision in the temporal field and an afferent pupil defect on the affected side. An MRI scan showed a pituitary mass

extending into the suprasellar cistern, displacing and stretching the optic chiasm (Figure 1). The vertical height of the lesion was 2.0 cm. Serum prolactin, measured several times, had risen by about 100 mU/L—no more than is usual in pregnancy. Other endocrine investigations were likewise normal so she was diagnosed as having a non-functioning pituitary tumour. The obstetric team were reluctant for her to undergo pituitary surgery in the early middle trimester since, if surgery precipitated labour, the fetus would not be viable.

The presentation in pregnancy was thought to be due to upward displacement of the tumour by physiological pituitary enlargement; thus dopamine agonist therapy might allow surgery to be deferred. She was started on bromocriptine (5 mg on day 1, 5 mg twice daily on day 2 and 5 mg three times daily thereafter). Five days after the beginning of treatment the visual symptoms had disappeared, and on a repeat MRI scan the optic chiasm was no longer compressed, although there was negligible change in the size of the tumour (Figure 2). The visual fields returned to normal. Bromocriptine therapy was continued and the pregnancy proceeded without incident; a healthy child was born at 39 weeks' gestation.

Postpartum, the patient was keen to breast-feed and elected to stop her bromocriptine. Within 2 days the visual field defect returned and she then underwent transphenoidal decompression of the tumour. On histological examination it proved to be a pituitary adenoma in which only about 5% of cells stained for prolactin and growth hormone. Postoperative endocrine assessment gave normal results except for hypoadrenalism. A year later she had an uncomplicated pregnancy.



Figure 1 MRI scan showing enlargement of the contents of pituitary fossa with compression of optic chiasm (arrow)

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Figure 2 MRI scan showing decompression of the optic chiasm after treatment with dopamine agonist (arrow)

**COMMENT**

We suspect that the beneficial effect of bromocriptine was achieved by an effect on the physiologically enlarged pituitary. A prolactinoma would be expected to respond to this agent<sup>3</sup>, but this patient's tumour was non-secretory; although 5% of cells stained for prolactin, serum prolactin concentrations were normal before and after pregnancy. 8–16% of non-secretory pituitary adenomas do respond to dopamine agonist therapy, so a direct effect cannot be ruled out; however, the rapidity of the therapeutic response makes this unlikely.

This is not the first report of successful use of bromocriptine to relieve compression of the optic chiasm by a non-secretory pituitary adenoma. In the previous cases, however, the effect was thought to be due to tumour shrinkage<sup>4,5</sup>. Grossman and colleagues suggest that in these circumstances dopamine agonists work by shrinkage of normal lactotrophs, and recommend only temporary use (for about 2 weeks) before surgery<sup>6</sup>. We could find no other cases where the physiological lactotroph hyperplasia of pregnancy had caused upward displacement of a clinically silent pituitary tumour.

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**Mesh repair of sacral hernia following sacrectomy**

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CLINICAL SECTION, 16 MAY 2002

Sacrectomy is used in the management of sacral or advanced pelvic tumours. A sacral hernia is a rare complication of the procedure.

**CASE HISTORY**

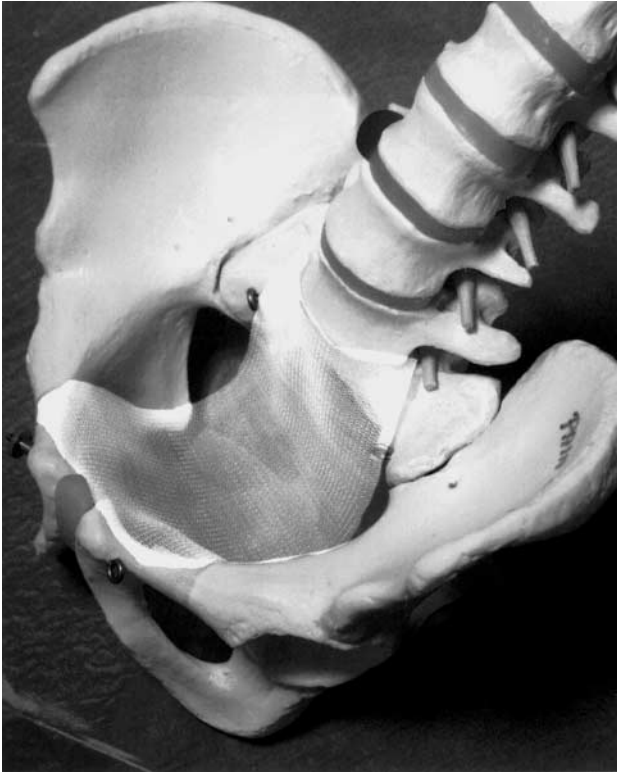
A man aged 68 underwent resection of a sacral chordoma. The tumour involved all sacral segments and nerve roots below S1, therefore the sacrum was divided at the S1/S2 junction. The anus and rectum were excised, and an end colostomy was fashioned in the left iliac fossa. Postoperatively he developed a sacral wound infection that required formal incision and drainage. He also had a prolonged ileus.

Four months later he reported sacral pain and swelling, and on examination he had a large sacral hernia. Initially he refused surgical repair, but 2 years later he developed gallstone pancreatitis and requested sacral hernia repair at the time of his open cholecystectomy. The hernia was repaired via a transabdominal approach. The hernial sac contained small intestine, which was reduced, and two sheets of polypropylene mesh were used to repair the hernial defect and reconstruct the pelvic floor (Figures 1 and 2). The caecum and omentum were used to cover the mesh, to lessen the risk of small-bowel adhesion. A single suction drain was used and prophylactic antibiotics were administered. There were no postoperative complications and at 2-year follow-up there was no evidence of hernial recurrence.

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*Figure 1* Representation of pelvic floor reconstruction: first layer of mesh placed in pelvis and sutured anteroposteriorly to the bony pelvis



*Figure 2* Second layer of mesh placed on top of, and sutured to, the first layer. Laterally, it was sutured to the iliac fascia

## COMMENT

Complications of sacrectomy include haemorrhage, infection, neurological injury affecting bowel, bladder, or lower limb function, and weakening of the pelvic ring thus impairing weightbearing and mobilization<sup>1</sup>. There have been few reported cases of sacral hernia repair<sup>1-4</sup>, although the incidence of symptomatic hernias occurring in the perineal region following abdominoperineal excisions of the rectum and pelvic exenterations has been reported as 1% and 3%, respectively<sup>5</sup>. Sacral hernias usually present with pain and swelling in the sacral region. They can cause bowel or bladder disturbance depending on the contents of the hernial sac. These symptoms are often improved by application of pressure over the hernia. It has been suggested that damage to the S2-S4 sacral nerves during sacrectomy predisposes to sacral hernia formation by denervating the levator ani, which normally supports the pelvic viscera<sup>1</sup>. In the present case there may have been additional predisposing factors—the postoperative wound infection, the raised intra-abdominal pressure secondary to paralytic ileus, and the patient's age.

There are three recognized surgical approaches for the repair of hernias occurring in the sacral and perineal region—the anterior (transabdominal), posterior (sacral or perineal) and combined anteroposterior approaches. The posterior approach is associated with less morbidity since the abdominal cavity is not entered, and is thus appropriate for high-risk patients and small hernias. However, the exposure is limited and may not allow examination for tumour recurrence or repair of an intra-abdominal structure injured during the procedure. The anterior approach allows full mobilization of the sac and its contents and facilitates the placement of a sheet of mesh under direct vision. However, it is associated with more postoperative morbidity and should perhaps be reserved for cases in which a laparotomy is required for other reasons<sup>5</sup>. A combined anteroposterior approach provides the best exposure and allows excision of redundant soft tissue once the hernia is reduced. However, most operators use this approach only in exceptional circumstances<sup>5</sup>.

Several surgical techniques have been used to repair the hernial defect. If the defect is small, the levator ani can be reapproximated with sutures, although this is usually difficult if the previous cancer resection has been adequate. Occasionally, the bladder or uterus can be sutured to the posterior pelvic wall to eradicate the defect. The pelvic floor can be reconstructed with prosthetic mesh, which is easy to use and can cover a deficiency of any size. If the defect is large, a myocutaneous flap involving gracilis, rectus abdominus or gluteus maximus can also be used. In the previous cases of sacral hernia repair, three groups used mesh and one sutured the uterus to the sacral remnant. No recurrences have been reported.

For reconstruction of the pelvic floor after sacrectomy, Localio *et al.*<sup>6</sup> advocated obliteration of the dead space by tight closure of the gluteus maximus. However, if the resection has been extensive, the residual soft tissue is often insufficient for adequate closure. In these circumstances Santora *et al.*<sup>1</sup> suggest that prosthetic mesh should be used prophylactically to reconstruct the pelvic floor and reduce the likelihood of sacral herniation<sup>1</sup>.

Symptomatic sacral hernia following sacrectomy is a rare but important complication, and during the initial surgical resection an attempt should be made to close the soft tissues adequately. Like other groups we have found that polypropylene mesh offers a simple, adaptable, and effective method of hernia repair and pelvic floor reconstruction.

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**Phacomatosis pigmentokeratolica**

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SECTION OF DERMATOLOGY

We report a 5-year-old girl with a rare and unusual variant of the epidermal naevus syndrome. There are a handful of cases previously reported.

**CASE HISTORY**

The patient was born at 36 weeks’ gestation after a pregnancy complicated by pre-eclampsia, weighing 2.5 kg. At birth she was noted to have three pigmented lesions on

the right forearm and a warty lesion on the right little finger. At 10 days of age erythema appeared on the right abdomen, arm and leg which gradually darkened and became slightly raised. A lentiginous area with raised pigmented lesions also appeared on the upper back. By the age of 18 months the pigmented lesions on the back, arm and hand had gradually extended. There was also an extensive epidermal naevus on the right of the body (Figure 1). The pigmented lesions on the upper back and the epidermal naevus on the lower back crossed the midline.

By the age of four years there were large melanocytic naevi in a speckled lentiginous pattern on the upper back, forearm and hand, composed of papules and nodules, some of which were reddish in colour. The epidermal naevus had sebaceous features and involved the right abdomen, arm and leg down to the foot. The configuration was in keeping with Blaschko’s lines<sup>1</sup>. On the medial aspect of the wrist the epidermal naevus coexisted with melanocytic naevus (Figure 2).

The child has severe epilepsy, which began as apnoeic attacks at the age of nine days and then manifested as ‘absences’, drop attacks, floppy episodes and partial complex seizures which can occur several times daily. She has been treated with sodium valproate, carbamazepine, lamotrigine and more recently carbamazepine with diazepam, all of which have been helpful. In addition, a mild right hemiparesis and developmental delay were diagnosed at the age of four months, both of which have virtually resolved. Investigations, including repeated electroencephalograms,



Figure 1 Age 18 months: epidermal naevus and pigmented lesions right forearm and back

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Figure 2 Age 4½ years: epidermal naevus right side of trunk and leg, with coexistence of epidermal naevus and melanocytic naevus right hand

video-telemetry, electrocardiogram, hearing test and chromosome analysis, were all normal. A recent MRI brain scan did show some enlargement of the left lateral ventricle, but this is not thought to be significant.

On histological examination, a skin biopsy from the right side of the abdomen revealed hyperkeratosis, papillomatosis and acanthosis. There were scattered mature sebaceous glands, hair follicles and eccrine glands in the epidermis, in keeping with a verrucous epidermal naevus with prominent sebaceous differentiation. Several pigmented lesions have been removed and proved to be benign melanocytic naevi, some with features of Spitz naevi.

Management has included surgical excision of some of the raised pigmented lesions on the right hand. Two laser treatments have been undertaken under general anaesthesia. Several of the pigmented lesions have been removed from the back and hand with the carbon dioxide laser, with variable results; some have already recurred but in other areas the cosmetic result is good. A test treatment on the epidermal naevus showed about 40% improvement but the parents decided against treatment of such a large area. The possible risk of malignant change within the pigmented areas is a concern.

## COMMENT

The epidermal naevus syndrome is the combination of a widespread epidermal naevus and other associated features including skeletal, neurological, eye and cardiac defects, developmental delay and precocious puberty. There are associated cutaneous abnormalities in at least one-third of patients: examples are woolly hair naevus<sup>2</sup>, naevus

comedonic, *café-au-lait* spots, hypopigmented patches, haemangiomas and melanocytic naevi.

Phacomatosis pigmentokeratolica is a rare and distinct variant of the epidermal naevus syndrome, first described by Happle *et al.*<sup>3</sup>. It comprises the association of an organoid (epidermal) naevus with sebaceous differentiation and a speckled lentiginous naevus<sup>4,5</sup> arranged in a checkerboard pattern. There are often other anomalies, most commonly neurological or skeletal, such as hemiatrophy, dysaesthesia and hyperhidrosis in a segmental pattern, mild mental retardation, seizures, deafness, ptosis and strabismus<sup>6</sup>.

The hypothesis of 'twin-spotting' has been proposed by Happle *et al.* to explain the coexistence of two separate and distinct naevi<sup>6</sup> in a segmental pattern, involving adjacent or corresponding regions of the body. Twin-spotting has been studied in plants, as well as in *Drosophila melanogaster*, and is based on the concept of somatic recombination due to an early post-zygotic mutational event. Two different autosomal recessive mutations must be located on the same chromosome pair, one on each homologous chromosome. The embryo is therefore double heterozygous. In early embryogenesis post-zygotic crossing-over and recombination results in two different homozygous populations of cells forming separate mosaic patterns. Another example of a similar process is phacomatosis pigmentovascularis—the combination of a widespread telangiectatic naevus and an extensive pigmentary naevus<sup>7</sup> which was described by Happle *et al.* before phacomatosis pigmentokeratolica.

Only a few cases of phacomatosis pigmentokeratolica have been recorded and we hope that other clinicians will recognize this unusual condition. There is a small but definite risk of malignant change within the pigmented lesions and the epidermal naevus in later life. Internal malignancies have been reported in the epidermal naevus syndrome, but none in phacomatosis pigmentokeratolica.

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## Antibiotic-resistant cellulitis acquired in Turkey

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When cellulitis proves unresponsive to the usual antibiotics, a history of travel may be relevant.

### CASE HISTORY

A woman of 70 sought advice from her general practitioner when her right lower leg had been painful and swollen for two weeks. A few days before the onset of symptoms, when she was on holiday in Turkey, she had sustained insect bites on that leg. Flucloxacillin and amoxycillin were prescribed, but the condition worsened and new lesions appeared on her left ankle. On referral to the dermatology department the patient had low-grade pyrexia and bilateral inguinal lymphadenopathy. Her right lower leg was hot and swollen with well-demarcated erythema and multiple pus-filled bullae (Figure 1). Similar lesions were present on the posterior aspect of the left ankle. White cell count was  $14.4 \times 10^9/L$  with 73% neutrophils; C reactive protein was 213 mg/L. Multiple skin swabs and pus aspirates were sent for microbiological examination. The bullous abscesses were incised and drained with partial relief of pain. High-dose intravenous benzylpenicillin and flucloxacillin were administered, with negligible benefit. X-rays of the chest and legs were normal.

The aspirates contained moderate numbers of pus cells but no evident organisms. After three days, however, *Nocardia asteroides*, with typical dry white colonies and an earthy odour, grew on blood agar from both skin swabs and pus aspirates. Blood cultures remained negative. Skin biopsies showed a dense mixed dermal inflammatory infiltrate with granulomata extending to subcutaneous fat (Figure 2). Gram and methanamine silver staining did not reveal any organisms; acid-fast staining was not done. The granulomatous histology and positive cultures from pus supported a diagnosis of primary cutaneous nocardiosis. She had not been in direct contact with soil and the infection was presumed to have been transmitted by the insect bites.

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Figure 1 Swelling of the right lower leg with well-demarcated erythema and multiple pus-filled bullae

Her immune function was normal. She was treated with cotrimoxazole 960 mg twice daily for three months and the lesions healed without recurrence.

### COMMENT

*Nocardia* spp. are soil saprophytes belonging to the order of Actinomycetales. The main species pathogenic in man are

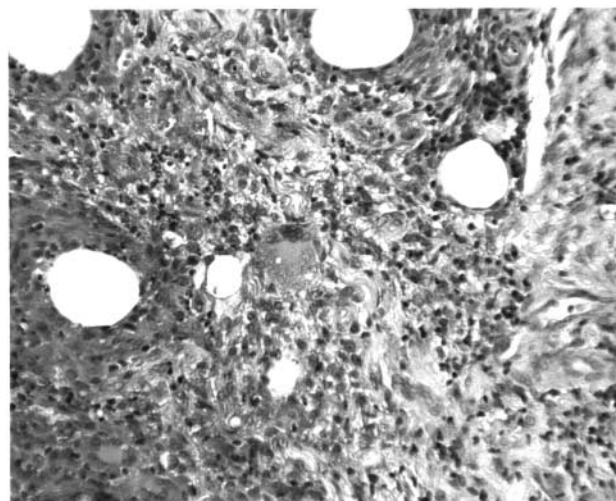


Figure 2 Skin biopsy of left ankle showing chronic granulomatous inflammation in dermis. A mixture of inflammatory cells is present including a foreign body giant cell in the centre ( $\times 25$ )

*N. asteroides* complex (*N. asteroides*, *N. nova*, *N. farcinica*) and *N. brasiliensis*. Nocardiosis is most commonly seen in Central and South America, North Africa and India. In France, a retrospective study suggested 150 to 250 new cases a year from 1987 to 1990<sup>1</sup>. The organism is a fine branching Gram-positive bacillus with an irregular beaded appearance. It is weakly acid fast but not alcohol fast and stains with modified Ziehl–Neelsen. It can take up to two weeks to grow under aerobic conditions on blood agar, Sabouraud's glucose agar or Löwenstein–Jensen medium. *Nocardia* spp. are readily overgrown by other organisms and may therefore be missed on culture.

Primary cutaneous nocardiosis has three distinct clinical presentations. First, a superficial infection can occur with cellulitis, abscess formation or multiple ulcers as in the case presented here. This presentation has not been reported often<sup>2</sup>, possibly because the condition is misdiagnosed as staphylococcal infection and on occasion resolves without specific antimicrobial therapy<sup>2</sup>. In rare circumstances, superficial cutaneous nocardia infection can disseminate to other organs. A second, lymphocutaneous, form of disease presents with clinical features similar to those of sporotrichosis; a cervicofacial subtype in childhood is recognized with cervical lymphadenopathy and nasolabial lesions, often with no preceding cutaneous injury. The third type of presentation is with an actinomycetoma or Madura foot—a chronic granulomatous mass with sinus formation. This must be differentiated from infection caused by *Actinomyces* spp. or fungi.

*N. brasiliensis* is the most common cause of primary cutaneous nocardiosis but overall only accounts for 10% of nocardia infection. Transmission is usually through a minor skin injury, including a cat scratch or thorn prick. The possibility of insect bites acting as a vector for the transmission of nocardia infection has been noted

previously<sup>3</sup>. Although it is unclear whether our patient had insect bites on both lower legs, the spread from the right to left leg can be explained by the direct skin to skin contamination. The incubation period of *Nocardia* spp. can vary from a week to several months. *N. asteroides* tends to be associated with pulmonary disease. From the lungs the infection can disseminate with widespread abscess formation<sup>4</sup>. Disseminated nocardiosis most commonly occurs in the setting of immunosuppression such as HIV infection, leukaemia or lymphoma and pre-existing chronic pulmonary disease.

The efficacy of antimicrobials in primary cutaneous nocardiosis is difficult to assess. The choice is often determined by their *in-vitro* activity<sup>5</sup>. Clinical experience and *in-vitro* activity favour co-trimoxazole. As an alternative, minocycline or a quinolone antibiotic may be effective but the sensitivity to such agents varies between species. Some of the intravenous agents such as amikacin, co-trimoxazole, cefotaxime and imipenem are reported to have synergistic *in-vitro* effects and may therefore be useful in combination for disseminated and severe cases.

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