GUIDELINES

Caesarean section: summary of updated NICE guidance

Maryam Gholitabar, Roz Ullman, David James, Malcolm Griffiths, on behalf of the Guideline Development Group

In England, rates of caesarean section have increased from 9% of births in 1980 to 24.8% in 2010. The indications for the procedure vary. Healthcare professionals have to provide evidence based information for women about the risks and benefits of both planned and unplanned caesarean section. To advise women appropriately they also need to be aware of specific indications for caesarean section, effective management to avoid unnecessary caesarean section and reduce morbidity from caesarean section, and birth after a caesarean section. This article summarises the most recent recommendations from the National Institute for Health and Clinical Excellence (NICE) on caesarean section.

Recommendations
NICE recommendations are based on systematic reviews of the best available evidence and explicit consideration of cost effectiveness. When minimal evidence is available, recommendations are based on the Guideline Development Group’s experience and opinion of what constitutes good practice. Evidence levels for the recommendations are in the full version of this article on bmj.com.

Possible reasons for caesarean section
Breech presentation (existing recommendations)
- For women who have an uncomplicated singleton breech pregnancy at 36 weeks’ gestation, offer external cephalic version (turning the baby). However, contraindications include women in labour, women with a uterine scar or uterine abnormality, fetal compromise, ruptured membranes, vaginal bleeding or certain medical conditions, such as severe pre-eclampsia and Rhesus isoimmunisation.
- For women with a singleton breech pregnancy at term for whom external cephalic version is contraindicated or has been unsuccessful, offer caesarean section because it reduces perinatal mortality and neonatal morbidity.

Morbidly adherent placenta
Women found antenatally to have morbidly adherent placenta (an abnormal adherence of the placenta to the uterine wall) will be advised to have a caesarean section.

Diagnosis of morbidly adherent placenta (new recommendation)
- If a colour flow Doppler ultrasound scan suggests a morbidly adherent placenta:
  - Discuss with the woman the improved accuracy of magnetic resonance imaging combined with ultrasonography, and explain what to expect during magnetic resonance imaging
  - Inform the woman that current experience suggests that magnetic resonance imaging is safe but that evidence is lacking about any long term risks to the baby

- Offer magnetic resonance imaging to improve the diagnostic accuracy and clarify the degree of invasion if acceptable to the woman.

Maternal request for caesarean section (new recommendations)
- When a woman requests a caesarean section explore, discuss, and record the specific reasons for the request.
- When a woman requests a caesarean section because she has anxiety about childbirth, offer referral to a healthcare professional with expertise in providing perinatal mental health support to help her address in a supportive manner.
- For women requesting a caesarean section, if after discussion and the offer of support (including perinatal mental health support for women with anxiety about childbirth) a vaginal birth is still not an acceptable option, offer a planned caesarean section.
- An obstetrician unwilling to perform a caesarean section should refer the woman to an obstetrician who will carry out the caesarean section.

Mother to child transmission of HIV (new recommendations)
- Do not offer a caesarean section on the grounds of HIV status to prevent mother to child transmission of HIV to (a) women receiving highly active antiretroviral therapy with a viral load of less than 50 copies per millilitre or (b) women receiving any antiretroviral therapy with a viral load of less than 50 copies per millilitre. Inform women that in these circumstances the risk of HIV transmission is the same for a caesarean section and a vaginal birth.
- Offer a caesarean section to women with HIV who (a) are not receiving any antiretroviral therapy, or (b) are receiving antiretroviral therapy and have a viral load of ≥400 copies per millilitre.

Planning mode of birth (new recommendation)
- Discuss with women the risks and benefits of caesarean section compared with vaginal birth (tables 1 and 2), taking into account their circumstances, concerns, priorities and plans for future pregnancies (including the risks of a morbidly adherent placenta with multiple caesarean sections).

Factors that reduce the likelihood of caesarean section (existing recommendations)
- Inform women that continuous support during labour from women with or without prior training reduces the likelihood of caesarean section.
- For women with an uncomplicated pregnancy offer induction of labour beyond 41 weeks because this reduces the risk of perinatal death and the likelihood of caesarean section.
### Effects around the time of birth

<table>
<thead>
<tr>
<th>Effect</th>
<th>Median Score</th>
<th>Median Score</th>
<th>Absolute Effect</th>
<th>Relative Effect</th>
<th>Evidence Quality</th>
</tr>
</thead>
<tbody>
<tr>
<td>Perineal and abdominal pain scores during birth*</td>
<td>Median score 1.0</td>
<td>Median score 7.3 (10.3%)</td>
<td>Pain score 6.3 lower</td>
<td>Not calculable</td>
<td>Very low</td>
</tr>
<tr>
<td>Perineal and abdominal pain scores 3 days postpartum</td>
<td>Median score 4.5</td>
<td>Median score 5.2 (10.3%)</td>
<td>Pain score 0.7 lower</td>
<td>Not calculable</td>
<td>Very low</td>
</tr>
<tr>
<td>Injury to vagina</td>
<td>0%</td>
<td>0.56% (14.7%)</td>
<td>6 fewer per 1000 (6 fewer to 2 fewer)</td>
<td>Not calculable</td>
<td>Very low</td>
</tr>
<tr>
<td>Early postpartum haemorrhage†</td>
<td>1.1%</td>
<td>6.0% (35%)</td>
<td>49 per 1000 (4 fewer to 56 fewer)</td>
<td>Odds ratio 0.23 (0.06 to 0.94)</td>
<td>Low</td>
</tr>
<tr>
<td>Obstetric shock</td>
<td>3.9%</td>
<td>6.2% (8.3%)</td>
<td>23 fewer per 1000 (35 fewer to 6 fewer)</td>
<td>Risk ratio 0.06 (0.4 to 0.9)</td>
<td>Very low</td>
</tr>
<tr>
<td>Obstetric shock</td>
<td>0.006%</td>
<td>0.018% (8.2%)</td>
<td>12 fewer per 100,000 (17 fewer to 0.1 fewer)</td>
<td>Risk ratio 0.33 (0.11 to 0.99)</td>
<td>Very low</td>
</tr>
</tbody>
</table>

### Studies suggesting reduction in effect after planned caesarean section

<table>
<thead>
<tr>
<th>Effect</th>
<th>Length of stay†</th>
<th>Hysterectomy done as a result of postpartum haemorrhage</th>
<th>Cardiac arrest</th>
<th>Median score</th>
<th>Median score</th>
<th>Odds ratio</th>
<th>P= /zero/.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Perineal and abdominal pain scores 3 months postpartum*</td>
<td>3.2 days</td>
<td>2.6 days (35%)</td>
<td>0.6 days longer</td>
<td>Mean difference 1.58 (1.27 to 2.17)</td>
<td>Low</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Injury to bladder or ureter</td>
<td>0%</td>
<td>0.14% (14.7%)</td>
<td>1 fewer per 1000 (2 fewer to 2 more)</td>
<td>Not calculable</td>
<td>Very low</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Injury to cervix</td>
<td>0%</td>
<td>0.28% (14.7%)</td>
<td>3 fewer per 1000 (3 fewer to 1 more)</td>
<td>Not calculable</td>
<td>Very low</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Iatrogenic surgical injury</td>
<td>0%</td>
<td>0.07% (14.7%)</td>
<td>7 fewer per 10 000 (10 fewer to 30 more)</td>
<td>Not calculable</td>
<td>Very low</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pulmonary embolism</td>
<td>0%</td>
<td>0.003% (14.7%)</td>
<td>2 fewer per 10 000 (2 fewer to 40 more)</td>
<td>Not calculable</td>
<td>Very low</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Wound infection†</td>
<td>0.01%</td>
<td>0.00% (35%)</td>
<td>1 more per 10 000</td>
<td>P=1.0</td>
<td>Low</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Intraoperative trauma</td>
<td>1.5%</td>
<td>0.9% (8.3%)</td>
<td>6 more per 1000 (1 fewer to 19 more)</td>
<td>Risk ratio 1.7 (0.9 to 3.2)</td>
<td>Very low</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Uterine rupture</td>
<td>0.1%</td>
<td>0.3% (8.3%)</td>
<td>1 fewer per 1000 (3 fewer to 7 more)</td>
<td>Risk ratio 0.5 (0.1 to 3.5)</td>
<td>Very low</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Assisted ventilation or intubation</td>
<td>0.01%</td>
<td>0.005% (8.2%)</td>
<td>7 more per 100 000 (0 fewer to 22 more)</td>
<td>Risk ratio 2.21 (0.99 to 4.90)</td>
<td>Very low</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Acute renal failure</td>
<td>0.004%</td>
<td>0.001% (8.2%)</td>
<td>2 more per 100 000 (9 fewer to 13 more)</td>
<td>Risk ratio 2.17 (0.58 to 8.14)</td>
<td>Very low</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

### Studies suggesting increase in effect after planned caesarean section

<table>
<thead>
<tr>
<th>Effect</th>
<th>Length of stay</th>
<th>Hysterectomy done as a result of postpartum haemorrhage</th>
<th>Cardiac arrest</th>
<th>Median score</th>
<th>Median score</th>
<th>Odds ratio</th>
<th>P= /zero/.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Perineal and abdominal pain scores 3 months postpartum*</td>
<td>3.96 days</td>
<td>2.56 days (8.2%)</td>
<td>1.4 days longer</td>
<td>Mean difference 1.58 (1.46 to 1.49)</td>
<td>Very low</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Injury to bladder or ureter</td>
<td>0.03%</td>
<td>0.01% (8.2%)</td>
<td>14 more per 100 000 (3 more to 33 more)</td>
<td>Risk ratio 2.51 (1.30 to 4.90)</td>
<td>Very low</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Injury to cervix</td>
<td>0.19%</td>
<td>0.03% (8.2%)</td>
<td>15 more per 10 000 (11.5 more to 19.5 more)</td>
<td>Risk ratio 4.91 (3.95 to 6.11)</td>
<td>Very low</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

### Studies finding no difference in effect

<table>
<thead>
<tr>
<th>Effect</th>
<th>Median score</th>
<th>Median score</th>
<th>Absolute effect</th>
<th>Relative effect</th>
<th>Evidence Quality</th>
</tr>
</thead>
<tbody>
<tr>
<td>Perineal and abdominal pain 4 months postpartum*</td>
<td>Median score 0.0</td>
<td>Median score 0.17</td>
<td>0.17 lower</td>
<td>Not calculable</td>
<td>Very low</td>
</tr>
<tr>
<td>Injury to bladder or ureter</td>
<td>0%</td>
<td>0.14% (14.7%)</td>
<td>1 fewer per 1000 (2 fewer to 2 more)</td>
<td>Not calculable</td>
<td>Very low</td>
</tr>
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<td>Injury to cervix</td>
<td>0%</td>
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<td>Iatrogenic surgical injury</td>
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<td>Not calculable</td>
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<tr>
<td>Pulmonary embolism</td>
<td>0%</td>
<td>0.003% (14.7%)</td>
<td>2 fewer per 10 000 (2 fewer to 40 more)</td>
<td>Not calculable</td>
<td>Very low</td>
</tr>
<tr>
<td>Wound infection†</td>
<td>0.01%</td>
<td>0.00% (35%)</td>
<td>1 more per 10 000</td>
<td>P=1.0</td>
<td>Low</td>
</tr>
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<td>Intraoperative trauma</td>
<td>1.5%</td>
<td>0.9% (8.3%)</td>
<td>6 more per 1000 (1 fewer to 19 more)</td>
<td>Risk ratio 1.7 (0.9 to 3.2)</td>
<td>Very low</td>
</tr>
<tr>
<td>Uterine rupture</td>
<td>0.02%</td>
<td>0.03% (8.2%)</td>
<td>1 fewer per 1000 (3 fewer to 7 more)</td>
<td>Risk ratio 0.5 (0.1 to 3.5)</td>
<td>Very low</td>
</tr>
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<td>Assisted ventilation or intubation</td>
<td>0.01%</td>
<td>0.005% (8.2%)</td>
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</tr>
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</table>

### Studies with conflicting findings

<table>
<thead>
<tr>
<th>Effect</th>
<th>Finding for planned vaginal birth (percentage of unplanned caesarean sections in planned vaginal birth group)</th>
<th>Absolute effect in planned caesarean section group (95% confidence interval)</th>
<th>Relative effect (95% confidence interval)</th>
<th>Evidence quality</th>
</tr>
</thead>
<tbody>
<tr>
<td>Maternal death†</td>
<td>9/737 (cases/controls); 13/49 (26.5%) of maternal deaths occurring in the planned vaginal birth group were women who gave birth by unplanned caesarean section</td>
<td>Not calculable</td>
<td>Odds ratio 2.28 (1.11 to 4.65)</td>
<td>Very low</td>
</tr>
<tr>
<td>Deep vein thrombosis†</td>
<td>0%</td>
<td>0.03% (14.7%)</td>
<td>0.7 fewer per 1000 (0.2 fewer to 4 more)</td>
<td>Not calculable</td>
</tr>
<tr>
<td>Blood transfusion†</td>
<td>1.7%</td>
<td>1.9% (35%)</td>
<td>2 fewer per 1000 (14 fewer to 34 more)</td>
<td>Odds ratio 0.87 (0.27 to 2.78)</td>
</tr>
<tr>
<td>Infection - wound and postpartum†</td>
<td>1.1%</td>
<td>0.8% (14.7%)</td>
<td>3 more per 100 000 (2 fewer to 11 more)</td>
<td>Risk ratio 1.36 (0.75 to 2.4)</td>
</tr>
<tr>
<td>Hysterectomy†</td>
<td>0.6%</td>
<td>0.21% (8.2%)</td>
<td>390 more per 100 000 (333 more to 464 more)</td>
<td>Risk ratio 2.85 (2.52 to 3.21)</td>
</tr>
<tr>
<td>Anaesthetic complications†</td>
<td>0.5%</td>
<td>0.21% (8.2%)</td>
<td>319 more per 100 000 (257 more to 389 more)</td>
<td>Risk ratio 2.5 (2.22 to 2.86)</td>
</tr>
</tbody>
</table>

*Score out of 10; higher scores indicate higher pain levels. †Findings reported from multiple studies.
Summary of effects on babies’ health of planned caesarean section compared with planned vaginal birth for women with an uncomplicated pregnancy and who have not had a previous caesarean section

<table>
<thead>
<tr>
<th>Effects around the time of birth</th>
<th>Finding for planned caesarean section</th>
<th>Finding for planned vaginal birth (percentage of unplanned caesarean sections in planned vaginal birth group)</th>
<th>Absolute effect in planned caesarean section group (95% confidence interval)</th>
<th>Relative effect (95% confidence interval)</th>
<th>Evidence quality</th>
</tr>
</thead>
<tbody>
<tr>
<td>Admission to neonatal intensive care unit</td>
<td>13.9%</td>
<td>6.3% (35%)</td>
<td>76 more per 1000 (31 more to 134 more)</td>
<td>Risk ratio 2.20 (1.4 to 3.18)</td>
<td>Low</td>
</tr>
</tbody>
</table>

Studies finding no difference in effect

- **Hypoxic-ischaemic encephalopathy (central nervous system depression, seizures, pH <7)**: 0.2% vs. 0.2% (14.7%).
- **Intracranial haemorrhage**: 0% vs. 0.01% (14.7%).
- **Neonatal respiratory morbidity**: 12.0% vs. 11.5% (14.7%).

Not calculable

Studies with conflicting findings

- **Neonatal mortality**: 0% vs. 0.1% (14.7%).
- **Apgar score at 5 mins <7**: 0% vs. 0.5% (14.7%).
- **Apgar score at 1 min**: 0.6% vs. 1.2% (35%).

*Findings reported from multiple studies.*

- Use a partogram (graphical representation of labour) with a four hour action line to monitor the progress of labour for women in spontaneous labour with an uncomplicated singleton pregnancy at term because such monitoring reduces the likelihood of caesarean section.
- Consultant obstetricians should be involved in the decision making for caesarean section because this reduces the likelihood of caesarean section.

**Anaesthesia for caesarean section (existing recommendations)**

- Provide women with information on the different types of analgesia available to them after a caesarean section so that they can be offered the analgesia best suited to their needs.
- Offer regional rather than general anaesthesia to women having a caesarean section (including women with a diagnosis of placenta praevia) because it is safer and results in less maternal and neonatal morbidity.
- Women who are having induction of regional anaesthesia for caesarean section should be cared for in theatre because this does not increase women’s anxiety.

**Timing of antibiotic administration (new recommendations)**

- Offer prophylactic antibiotics to women having a caesarean section to reduce the risk of postoperative infections. Choose antibiotics effective against endometritis and urinary tract and wound infections, which occur in about 8% of women who have had a caesarean section.
- Offer prophylactic antibiotics before skin incision. Inform women that this reduces the risk of maternal infection more than prophylactic antibiotics given after skin incision and that research has shown no effect on babies.
- Do not use co-amoxiclav when giving antibiotics before skin incision because of a proved increased risk of necrotising enterocolitis in babies when used in women presenting with preterm labour.

**Pregnancy and childbirth after caesarean section (new recommendations)**

- Inform women who have had up to and including four caesarean sections that the risk of fever, bladder injuries, and surgical injuries does not vary with planned mode of birth and that the risk of uterine rupture, although higher for planned vaginal birth, is rare.
- While women are in hospital after having a caesarean section, give them the opportunity to discuss with healthcare professionals the reasons for the caesarean section and provide both verbal and printed information about birth options for any future pregnancies. If the woman prefers, provide the information at a later date.

**Overcoming barriers**

Caesarean section rates in the United Kingdom, as in all developed countries, have risen over recent years, although the absolute rates vary. Possible reasons for the rise include changes in sociodemographic factors, clinical practices (including a repeat elective caesarean section in women who have had the procedure before), and the attitudes of professionals and women. Implementing the guideline by providing women with evidence based information on the risks and benefits of planned vaginal birth and caesarean section will promote effective communication and empower them to make informed decisions.

Some service providers and commissioners may feel that the guideline will result in a large increase in caesarean births on maternal request. However, the recommendations show that the evidence for vaginal birth for such women is robust.

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Table 1 (left) Summary of effects on women’s health of planned caesarean section compared with planned vaginal birth for women with an uncomplicated pregnancy and who have not had a previous caesarean section

Table 2 Summary of effects on babies’ health of planned caesarean section compared with planned vaginal birth for women with an uncomplicated pregnancy and who have not had a previous caesarean section

<table>
<thead>
<tr>
<th>Effects around the time of birth</th>
<th>Finding for planned caesarean section</th>
<th>Finding for planned vaginal birth (percentage of unplanned caesarean sections in planned vaginal birth group)</th>
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- **Apgar score at 5 mins <7**: 0% vs. 0.5% (14.7%).
- **Apgar score at 1 min**: 0.6% vs. 1.2% (35%).

*Findings reported from multiple studies.*
A PATIENT’S JOURNEY

Amyloidosis

Malvyn Benjamin,1 Simon Gibbs2

After several months of renal and respiratory investigation, Malvyn Benjamin was diagnosed with amyloidosis. Here he describes his experience of the condition and its treatment.

I was diagnosed with amyloidosis in June 2010 at the age of 73. The sequence of events was interesting. I had been attending the hospital for many months, seeing consultants in the nephrology and respiratory departments, unfortunately to no effect.

For several months my ability to walk long distances had been impaired by extreme breathlessness: on a journey of 10 to 15 minutes’ duration I would have to stop 20 or 30 times to catch my breath. At one point, after such a walk, I collapsed in a synagogue—which was actually a good place to collapse, to 15 minutes’ duration I would have to stop 20 or 30 times to catch my breath. At one point, after such a walk, I collapsed in a synagogue—which was actually a good place to collapse, as I was surrounded by many doctors. After routine blood and urine tests at my general practitioner’s surgery, my doctor telephoned me to say that she had spotted something and made an urgent appointment at the haematology department in my local hospital.

There, I was put through a battery of tests and was told my results would be sent to the National Amyloidosis Centre for examination. Of course, I had never heard of amyloidosis. Before going to the centre, I had a bone marrow test—not the most pleasant experience—and later, at the centre, I had various other tests, including an electrocardiogram and a full body scan.

When the results of the tests and scan came through I was told by the doctor at the centre that I had amyloidosis and that if I did not receive treatment for it I would not last beyond the end of the year.

My condition involves amyloid deposits in my kidneys, which were working at 20% of the normal rate, and in my

The guidance may reduce the number of unnecessary caesarean sections and the associated morbidity, benefiting women and babies. The aim is for all caesarean sections to be appropriate.

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Provenance and peer review: Commissioned; not externally peer reviewed.


Amyloidosis is a multisystem disease, characterised by the misfolding of proteins into highly organised, insoluble amyloid fibrils that deposit in tissues. It is estimated that up to one in 1500 people are affected by amyloidosis in the UK. Without treatment, amyloid fibrils accumulate and lead to organ impairment, failure, and ultimately death. Organ involvement most commonly includes the kidneys, heart, nerves, and soft tissues.

Diagnosis is made by biopsy of the affected organ (for example, kidney) or by a screening biopsy such as a deep rectal biopsy involving submucosa by flexible sigmoidoscopy. Serum amyloid P scintigraphy, which specifically images visceral amyloid deposits, is available at the National Amyloidosis Centre and provides information about the amyloid load throughout the body. Repeating this scintigraphy every six to 12 months is an excellent means of tracking regression (or progression) of amyloidosis post treatment. There are four main categories for systemic amyloidosis.

- AL (light chain) amyloidosis—associated with a plasma cell dyscrasia or myeloma. This is the most common and lethal form of amyloidosis. It requires urgent diagnosis and treatment. Chemotherapy—typically cyclophosphamide and dexamethasone with bortezomib or thalidomide—suppresses production of the amyloidogenic monoclonal serum free light chain. The aim of treatment is to suppress the clone as rapidly and deeply as possible while limiting toxic effects. A 90% or greater suppression of the monoclonal serum free light chain component can result in substantial improvements in organ function and overall survival.

- AA (serum amyloid A protein) amyloidosis—associated with chronic inflammation, for example, rheumatoid arthritis, Crohn’s disease, bronchiectasis and inherited liver syndromes such as familial Mediterranean fever. The precursor protein for this amyloid type is serum amyloid A protein, an inflammatory protein similar to C-reactive protein. Chronic raised concentrations of serum amyloid A over many years can result in this type of amyloidosis, typically involving the kidneys, leading to substantial renal impairment or failure with nephrotic syndrome. Treatment is suppression of the underlying cause of inflammation, such as with anti-tumour necrosis factor drugs in rheumatoid arthritis. With adequate control of inflammation (serum amyloid A concentrations persistently <10mg/L), prognosis can be excellent.

- Hereditary amyloidosis—there are several types of hereditary amyloid diseases, and these can mimic systemic AL amyloidosis. A family history can often be absent, and chemotherapy is ineffective. Screening for amyloidogenic genetic mutations, where relevant, is part of the routine assessment at the National Amyloidosis Centre, after patient counselling and consent. Solid organ transplantation can be used in some cases to replace failed organ function.

- Senile systemic amyloidosis—amyloid fibrils from misfolded wild-type (“normal”) transthyretin protein deposit in the carpal tunnels and cardiac tissue, leading to a restrictive cardiomyopathy. This condition is most commonly seen in older, white men. Treatment is with diuretics and standard anti-failure medication.

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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.

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• Facial disfigurement (BMJ 2011;343:d5203)
• Living with alkaptonuria (BMJ 2011;343:d5155)
• Dementia with cardiac problems (BMJ 2011;343:d4278)
• Destined to die (BMJ 2011;343:d3625)
A DOCTOR’S PERSPECTIVE

Malvyn has systemic AL amyloidosis, confirmed on serum amyloid P scintigraphy and bone marrow biopsy. His disease led to a substantial restrictive cardiomyopathy, diastolic dysfunction, nephrotic syndrome, and renal impairment. Unfortunately, his story of delayed diagnosis is not uncommon. Presenting symptoms can often be vague—such as lethargy, dyspnoea, fluid retention, dizziness, weight loss, and disturbed bowel function. More suggestive clinical signs can include periorbital bruising (“raccoon eyes”) and macroglossia. Any medical professional reviewing a patient with a restrictive cardiomyopathy, significant proteinuria, or a progressive peripheral or autonomic neuropathy should have systemic amyloidosis in their list of differential diagnoses. Malvyn achieved a 65% clonal response with four cycles of chemotherapy (cyclophosphamide and dexamethasone with thalidomide) but with considerable toxic effects, particularly fluid retention. The decision was made to replace the thalidomide component with bortezomib. Malvyn tolerated the bortezomib well, and his clonal response improved to 85%. This led to an improvement in Malvyn’s energy levels, a reduction in proteinuria and dyspnoea, and stabilisation of his renal impairment and cardiomyopathy. Balancing treatment efficacy with toxicity is a constant challenge. However, despite initial significant side effects, Malvyn’s treatment has improved both his quality of life and his lifespan. His fighting spirit, his faith, and the support of his family were a huge help. Amyloidosis is still a challenging and incurable disease, but with increasing understanding and treatment options, patients are surviving longer. We are developing new treatments that target amyloid deposits directly at the UCL Centre for Amyloidosis and acute phase proteins in a collaborative programme with GSK. Preliminary clinical trials at the National Amyloidosis Centre will commence shortly.

Simon Gibbs

USEFUL RESOURCES

National Amyloidosis Centre, UK (www.ucl.ac.uk/medicine/amyloidosis/nac)—Any patient with symptoms or signs suggestive of amyloidosis can be discussed with or referred to the National Amyloidosis Centre. Patients undergo serum amyloid P scintigraphy, echocardiography, and relevant blood, urine, and genetic testing as part of their clinical assessment. Further tests such as cardiac magnetic resonance imaging, $^{99m}$Tc-3,3-diphosphono-1,2-propanodicarboxylic acid scintigraphy, or nerve function tests are also arranged if required. Any biopsies will undergo confirmatory histological review, including immunospecific staining to confirm amyloid type, and a detailed explanatory diagnostic and management advice report is sent to the referring doctors, the general practitioner, and the patient. Patients are followed up every three to 12 months depending on their individual needs, and a shared model of care with local physicians is encouraged. Myeloma UK (www.myeloma.org.uk)—registered charity offering a broad and innovative range of services covering every aspect of myeloma including systemic AL amyloidosis, from information and support to improving standards of treatment and care through research, education, campaigning, and raising awareness.

heart. The heart wall was thickened and it is this that causes my breathlessness. There are also amyloid deposits in my liver.

When I went back to the haematology department, the consultant prescribed a regimen of pills for me to take. I was taking between 20 and 30 pills a day. My wonderful daughters—I have five—drew up a daily schedule for me with a cabinet into which the pills were placed for me to take daily.

This continued for several months, but it became apparent both to me and to my doctors that the pills were not having the desired effect. Eventually, in October 2010, it was decided that I should have an intensive course of chemotherapy with bortezomib.

I cannot say that I have taken easily to the chemotherapy. I generally feel pretty rotten after it has been administered, and it leaves me totally exhausted. I have lost my appetite and have the greatest difficulty in sleeping. I may sleep for an hour or two but then be awake for the rest of the night. I have become an expert on the BBC World Service, to which I listen while others sleep.

In November 2010 one of my daughters got married. There was considerable doubt among my family and friends as to whether I would actually make it to the wedding. It was agreed that my chemotherapy should be arranged so that I could take a week off for the wedding, but to no avail; in the event, chemotherapy was scheduled for the wedding day. However, I managed to put it off, so I was present when my daughter got married. It was a tremendous lift for me, and this feeling lasted for some time. The lesson I drew from it is that it is always a bonus to have something to aim for outside the medical situation, and the wedding sustained me.

I have now completed five cycles of chemotherapy. I am told there has been a dramatic improvement in my condition, and it has been decided to bring the chemotherapy to an end. Hopefully my amyloidosis will stabilise. If it does not, other options will be considered.

I have every confidence in my haematologist and an absolutely wonderful nurse, and my new nephrologist. I see the haematologist every four or five weeks and the nephrologist every two months. I also have checks every six months at the National Amyloidosis Centre. I guess it will be some time before I cease to have these appointments.

However, since I stopped chemotherapy, I no longer have to suffer the very unpleasant side effects of diarrhoea and constipation. Indeed, there has been a real improvement in my quality of life so that I am now able to sleep through the night and my appetite has returned. I can walk further than before, and the breathlessness, although still present, is no longer the issue that it was before I was diagnosed and before chemotherapy.

When I was originally diagnosed, my family faced the real possibility of losing their husband and father. Naturally, stress levels increased for all of us. I think we all decided to live each day as it comes, not least because the alternative—to concentrate on my imminent mortality—was not to be contemplated. I myself certainly resolved to put that to the back of my mind. I am grateful to my general practitioner, who set the process in motion; she is a compassionate and caring doctor. And I consider myself very fortunate, despite all the anguish my condition has caused. Had this happened 15 or 20 years ago I would not have been around to tell the tale. It is also a happy coincidence that the National Amyloidosis Centre is located in my local hospital, the Royal Free Hospital in London.

For all that, I do still harbour a strong resentment that so many health professionals seem completely ignorant of amyloidosis. I accept that it is a relatively rare condition, with only 500-600 new cases diagnosed in the UK each year; however, the disorder is almost certainly underdiagnosed. My concern is that it simply never occurred to the consultants in the nephrology and respiratory departments to consider the possibility of amyloidosis. It just was not on their radar. Indeed the nephrology consultant said he did not understand why my kidney function was declining and said, “I will see you in a year.” My response was that I could be dead in a year! My respiratory consultant suggested I have a lung function test and the result was normal. Once again an alternative cause was never considered.

I am just one patient. Others may have different experiences, but we all have one factor in common: there is no cure for amyloidosis. It is potentially fatal. But research is currently being undertaken to try to find a cure. I hope and pray that it will not be too late for me. In the meantime, however, the doctors manage the condition, so far with a fair degree of success. They are a very devoted and committed group.

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