

half of more than 200 patients on whom doctors were asked to complete personal medical attendants' reports could not recall having given their permission. A third objected to questions on sexual habits, and over half expected their doctors to withhold sensitive information. This report should help general practitioners when preparing reports for insurance companies, and it suggests that they should make sure that their patients know that they are completing them: they might also discuss with the patients whether they are happy for certain information to be released.

Insurance companies should consider carefully the questions they ask so that doctors' relationships with their patients are not harmed. For instance, a question such as "Is there anything about the patient's lifestyle that makes it likely that the patient will get a sexually transmitted disease?" puts

an improper strain on the professional relationship. Such questions should be directed at the patient; doctors should be expected to report medical fact not conjecture.

The relation between insurance companies and the proposer's general practitioner is important to nurture as together they may help the proposer to achieve a just contract. Great care and goodwill are needed in negotiating between the two groups to ensure a happy relationship and to reassure patients that their doctors will disclose only medical facts for which they have given informed consent.

JOHN D BEALE

General Practitioner,
Ramsgate, Kent CT11 9NB

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Causes of cataract

Age, sugars, and probably ultraviolet B radiation

Around the world about 17 million people are blind because of cataract.¹ Although cataract is treatable by operation, increasingly the resources of poor countries are outstripped by the growing demand.² In rich countries an increasing proportion of resources is consumed by treating cataracts. In England and Wales operations for cataracts increased by almost two thirds in the decade up to 1985,³ and there was an increase of 177% in the decade before 1976 in the United States,⁴ where one million cataract operations were carried out in 1987. Clearly, prevention of cataract should be emphasised, but unfortunately the cause of the commonest form of cataract—that related to aging—remains an enigma.

There are many causes of cataract, which is the common response to a physical, mechanical, or chemical insult.⁵ The transparency of the lens of the eye depends on a unique arrangement of tightly packed fibres, which in turn rely on a certain protein structure; and the lens is isolated in its special environment by a capsule and epithelium. Hence damage to the capsule, epithelium, or the constituent fibres of the lens may all lead to the formation of a cataract, and the injury may be cumulative over many years.

The most studied cataracts are those that may be caused by high concentrations of various sugars.⁶ The best understood form of cataract occurs in galactosaemia, which results from a lack of the enzyme galactose-1-phosphate uridyltransferase or galactokinase. The galactose that accumulates is converted to galactitol in the presence of aldose reductase and leads to increased osmotic pressure and hydration as galactitol does not diffuse out of the capsule. An aldose reductase inhibitor would prevent the conversion of galactose to galactitol and has been shown to prevent and reverse early cataract when instilled topically into the eyes of galactosaemic rats.⁷

In diabetics under 60 the prevalence of cataract is three or four times that in the normal population,⁸ and the cataracts may develop similarly to those in patients with galactosaemia—yet whereas the evidence in animal studies is suggestive the case in humans is less convincing.⁹ An alternative hypothesis is that non-enzymatic glycosylation occurs¹⁰: this explains both the increased pigmentation and protein aggregation in the lens and accords with the finding of a blood glucose concentration that is higher in patients with cataracts than in controls.¹¹

Much more important than the effect of sugars in causing

cataract is the potential effect of electromagnetic radiation. The damaging effects on the lens of ionising, microwave, and infrared radiation are well known,¹² but hitherto the effects of ultraviolet radiation from the sun have been uncertain. Most of the ultraviolet radiation is filtered out by the cornea, and only wavelengths of 295 nm or greater pass through. But the lens absorbs nearly all the ultraviolet from 295 nm to 400 nm, and the high prevalence of cataract in countries with hot climates has meant that the harmful effect of ultraviolet radiation has long been suspected.¹³

Some epidemiological data already support the hypothesis that absorbing ultraviolet radiation from sunlight is an important risk factor in forming cataracts.¹⁴ The hypothesis fits the observations of increased yellowing of the lens nucleus with aging¹² and that exposure to ultraviolet radiation leads to the formation of chromophores from proteins containing tryptophan, thus increasing the potential of the nucleus to absorb more radiation.¹⁶ The precise mechanism is still disputed but may depend on the oxidation of free radical scavengers setting in train reactions that lead to protein aggregation.¹⁷

Although this hypothesis fits the changes that occur in the nucleus, it does not match the observation that age related cataracts commonly affect the outer cortex. Furthermore, epidemiological data from India suggested that the prevalence of cataract was higher in the cloudier plains of Punjab than in the sunnier hills of the Himalayas.¹⁸ A later study repeated in the same region found a correlation between cataract and the concomitants of poverty, notably poor nutrition and hygiene.¹⁹ In such cases a powerful risk factor may be dehydration from serious diarrhoea, which may increase blood urea concentrations and thus make cyanate more available for carbamylation of lens protein.²⁰ Diarrhoea was an important risk factor in a study in Madhya Pradesh²¹ and (somewhat surprisingly) in Oxford.²²

More recently a tightly controlled study on watermen in the Chesapeake Bay showed correlation between cataract and prolonged exposure to sunlight²³; exposure to ultraviolet B radiation (295-320 nm) was important whereas that to ultraviolet A radiation (320-400 nm) was fairly unimportant, and the positive relation was with cortical rather than nuclear cataract.

Although more evidence is accumulating to support the

assertion that sunlight is important in causing cataract, sunlight is likely to be one of the many causative factors.¹¹ But at a time when there is ample evidence to suggest thinning of the protective ozone layer in the outer atmosphere²⁴ it makes sense to advise patients to minimise their exposure to ultraviolet radiation to protect not only the lens but also the retina and the skin.²⁵

HUNG CHENG

Consultant Ophthalmologist,
Eye Hospital,
Oxford OX2 6AN

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Managing biliary atresia

Referral before 6 weeks is vital

Before Kasai and Suzuki described portoenterostomy 30 years ago the mortality in biliary atresia was almost 100%.¹ Most children died in infancy. Now high survival rates may be achieved—but only if cholestasis is diagnosed and the affected child referred early.

Biliary atresia is not a congenital malformation but an acquired progressive disease of the biliary system. Its cause is unknown. Most infants develop cholestasis shortly after birth, but it may start weeks later. Histological studies of the biliary system excised at operation have illustrated the acquired inflammatory process.^{2,3}

All babies developing cholestasis need hospital investigation so that the nature of the jaundice can be defined; genetic, metabolic, and infective causes of intrahepatic cholestasis can be identified; and babies with extrahepatic biliary obstruction who need an operation can be recognised.⁴ The severity of cholestasis is assessed by radiopharmaceutical tests,⁵ which help to identify infants with extrahepatic obstruction. Liver biopsy is also indicated. Although the result of no single investigation is completely reliable, the site of cholestasis is identified in over 95% of cases. Investigations will diagnose liver disease that responds to specific treatment and prevent unnecessary and possibly harmful operations on infants with intrahepatic cholestasis.

Most previously untreatable infants with biliary atresia now obtain satisfactory biliary drainage after hepatic portoenterostomy. Some patients have lived beyond the age of 20 without features of liver disease, and the treatment continues to improve—partly because of improvements in the skill of surgeons^{6,7} but also because of modifications of the original procedure. Some modifications—for example, using microsurgical techniques⁸ and operating again if bile drainage remains impaired^{9,10}—increase the prospect of satisfactory bile drainage postoperatively. Other modifications reduce the risk of ascending cholangitis,¹¹⁻¹³ a common complication that may cause further deterioration in liver function.¹⁴

There is no agreement on which is the best operation.

Although more complicated procedures and repeat operations may increase the bile flow, they also reduce the prospect of successful liver transplantation. Biliary atresia with advanced liver disease is the commonest indication for liver transplantation in children,¹⁵ and previous biliary operations increase the risk of complications such as bleeding during hepatectomy. The poor postoperative results and the unavailability of donor livers mean that liver transplantation will not replace portoenterostomy as the initial operation for biliary atresia.¹⁶ Japan, where so many developments have originated, does not have a programme of liver transplantation.

Bile drainage and prognosis seem to be improved if patients who have had portoenterostomy are given total parenteral nutrition,¹⁷ corticosteroids,¹⁸ antibiotics to reduce the risk of early cholangitis,¹⁹ and possibly hormones and other therapeutic agents to encourage bile flow.^{20,21} The age at operation is crucial in determining the outcome of portoenterostomy.^{6,7,9,16,19,22} Infants operated on before 2 months have a 90% chance of eventual satisfactory bile drainage¹⁷; beyond this age the success rate rapidly falls and is nil by 5 months.¹⁷ Some infants with cholestasis are referred to specialist centres far too late. Two weeks are needed for the preoperative investigations, and infants must therefore be referred before the age of 6 weeks. Jaundiced infants are likely to have cholestasis if jaundice is still present at 6 weeks old, if the urine never becomes colourless, or if the stools lack the normal yellow or green pigment.²² Such infants must be referred to specialist centres.

R NELSON

Consultant Paediatrician,
Department of Child Health,
Royal Victoria Infirmary,
Newcastle upon Tyne NE1 4LP

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