Comment

These four patients show the wide range of syndromes with which cervical subluxation may present and emphasise the importance of considering the neck as a cause of neurological deficit in patients with rheumatoid arthritis. The Brown-Séquard syndrome has been reported only once before in association with altantoaxial subluxation due to rheumatoid arthritis.[3] Proprioception and vibration sense were retained in the first patient, reflecting asymmetric anterior compression of the spinal cord by the subluxed odontoid peg. Sensory deficit in a glove and stocking distribution in the patient in case 2 could have been confused with the peripheral neuropathy of rheumatoid arthritis, but it cleared with cervical spine traction and fusion, suggesting that it was due to cord compression. A similar case was reported by Marks and Sharp.[4] The presenting symptom of dyspnoea was most unusual; only on further questioning and physical examination did the more usual features of cervical cord compression become evident. "Drop attacks" in association with cervical subluxation are ominous and have been described before in association with atlantoaxial subluxation; our fourth case serves as a reminder. The attacks are usually attributed to brainstem ischaemia secondary to vertebral artery compression, which may also explain the respiratory and cardiovascular disturbances in these patients.[2] Myelography was not performed in the patients in cases 3 and 4. Myelography is advisable in all such cases because the rheumatoid changes may not be confined to the atlantoaxial joint, and the major source of compression may be at a lower level.[5]

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For Debate . . .

Impact of maternal serum alpha-fetoprotein screening on antenatal diagnosis

DAVID J H BROCK

Abstract

An analysis has been made of indications for amniocentesis in the Edinburgh area from 1979 to 1981. About 5% of all mothers underwent the procedure. Among 2137 amniocenteses, 37% were performed on mothers 35 years old or more, and 30% on patients with raised serum α-fetoprotein. The total number of amniocenteses and the categories have been stable for the past three years. As a result of amniocentesis 104 pregnancies were terminated, 66 of which (63%) followed a raised maternal serum α-fetoprotein indication, while only 10 (9·6%) were in mothers aged 35 or more. There were a further 12 terminations based on raised serum α-fetoprotein but where no amniocentesis had been thought necessary. Even when figures for anencephaly are excluded from the analysis, maternal serum α-fetoprotein screening was responsible for detecting 35 out of 63 (56%) abnormal fetuses. This constitutes a strong case for the continuation of α-fetoprotein screening programmes.

Department of Human Genetics, University of Edinburgh, Western General Hospital, Edinburgh EH4 2XU
DAVID J H BROCK, PHD, MRCPATH, reader in human genetics

Introduction

One of the aims of early antenatal diagnosis is the reassurance of parents who have an increased risk of bearing a child with a serious birth abnormality. The other aim is to detect fetal disorders early enough to allow the possibility of termination of pregnancy. Though reassurance is important in counselling and obstetric management, the effectiveness of antenatal diagnosis must in the end be judged by its ability to reduce the birth prevalence of serious disorders. One way of judging effectiveness is to examine current indications for amniocentesis and the frequency with which they allow abnormal fetuses to be detected. In this report I have analysed experience in one part of Scotland over the past three years.

Materials and methods

Antenatal diagnosis in Edinburgh is carried out in five separate laboratories. All amniotic fluids are referred to my laboratory for α-fetoprotein analysis, and it has therefore been possible to construct and maintain a central register of indications for amniocentesis and outcomes of pregnancy for the whole region. I have analysed the indications for amniocentesis and the terminations of pregnancy resulting from laboratory determinations for the years 1979-81. The three most recent years are chosen since the increasing demands on diagnostic services seen in the early and mid-1970s appear to have reached a plateau. The analysis is restricted to samples taken in south-

east Scotland (Lothians, Borders, and Fife), and excludes all those sent from outside this region.

Attempts have been made to ascertain the outcome of all pregnancies where amniocentesis has been performed. This has been only partially successful. The object of this report, however, is to assess the effectiveness of antenatal diagnosis in terms of the relation between indications for amniocentesis and fetal disorders detected. Though it is possible that some missed diagnoses may not have been reported, it is my experience that these rare events are usually promptly communicated to the responsible laboratory.

Results

INDICATIONS FOR AMNIOCENTESIS

The indications for amniocentesis have been divided into six categories (table I).

Raised maternal serum α-fetoprotein denotes those individuals who have been screened and found to have two sequential values above the 95th percentile of the normal range (or occasionally one very high value).

Previous neural tube defect includes mothers who have had an earlier pregnancy (or pregnancies) where the outcome was spina bifida or anencephaly. Parents who themselves suffer from spina bifida have been included in this category, since their risk of bearing a child with a neural tube defect is also 4% to 5%.

Family history of neural tube defect comprises a group where the brothers or sisters of the index parents have had offspring with neural tube defects, and where amniocentesis and antenatal diagnosis is usually offered.

Maternal age consists of mothers aged 35 or more.

Family history of chromosome abnormality includes mothers who have had a child with a chromosome abnormality as well as parents who are translocation carriers.

Other—The category labelled *other* is somewhat miscellaneous and includes maternal anxiety, as well as mothers with abnormal ultrasonar scans, histories of recurrent abortion, and previous children with inborn errors of metabolism. More than half of this group were made up by mothers under the age of 35 who sought amniocentesis for reassurance.

On many occasions when there was more than one indication for amniocentesis the pregnancy was assigned to the category with the highest risk. Thus a 40-year-old mother with a child with spina bifida was regarded as having a "previous neural tube defect" indication rather than one related to maternal age. Pregnancies were only placed in the "raised maternal serum α-fetoprotein" group, however, when no decisions on offering amniocentesis had been made before the results of the serum α-fetoprotein determination were known.

Terminations of pregnancy—Terminations of pregnancy are related to the indications for amniocentesis in table II. The data exclude terminations based on raised maternal serum α-fetoprotein and ultrasonar scan but where no amniocentesis was performed. My records of this last group are necessarily incomplete, though terminations of 11 fetuses with anencephaly and one with spina bifida in the three-year period have been discovered. These extra 12 neural tube defects have been included in table III, which shows terminations of anencephaly and spina bifida by individual years.

TABLE I—*Indications for amniocentesis, south-east Scotland, 1979-81*

Year	No of amniocenteses	Indications for amniocentesis (% of total)					
		Raised maternal serum α-fetoprotein	Previous neural tube defect	Family history of neural tube defect	Maternal age	Family history of chromosome disorder	Other
1979	690	220 (32)	73 (11)	29 (4)	235 (34)	56 (8)	77 (11)
1980	740	219 (30)	62 (8)	38 (5)	285 (39)	68 (9)	68 (9)
1981	707	198 (28)	74 (10)	21 (3)	275 (39)	57 (8)	82 (12)
All	2137	637 (30)	209 (10)	88 (4)	795 (37)	181 (8)	227 (11)

TABLE II—*Terminations of pregnancy related to indications for amniocentesis (1979-81)*

Indications for amniocentesis	No	Terminations of pregnancy		
		No (%)	Outcomes	No
Raised maternal serum α-fetoprotein	637	66 (10·4)	Anencephaly	32
			Spina bifida	24
			Exomphalos	5
			Encephalocele	2
			Turner's syndrome	1
			Fetus papyraceous	1
			Congenital skin defect	1
Previous neural tube defect	209	11 (5·3)	Anencephaly	4
			Spina bifida	3
			Meckel's syndrome	2
			Trisomy 21	2
Family history of neural tube defect	88	2 (2·3)	Trisomy 21	1
			46, XY, 4p−	1
Maternal age	795	10 (1·3)	Trisomy 21	7
			Trisomy 18	1
			Spina bifida	2
Family history of chromosome abnormality	181	2 (1·1)	Trisomy 21	1
			Trisomy 18	1
Other	227	13 (5·7)	Anencephaly	6
			Other*	7
All	2137	104 (4·9)	Anencephaly	42
			Spina bifida	29
			Exomphalos	5
			Trisomy 21	11
			Other	17

*Three male fetuses at risk for Duchenne muscular dystrophy; one metachromatic leucodystrophy; one Gaucher's disease; one 46, XY, 1p+; one unspecified chromosome abnormality.

TABLE III—*Terminations of neural tube defect fetuses by year*

Year	Terminations as a result of raised maternal serum α-fetoprotein		Terminations on other indications		All terminations
	Anencephaly	Spina bifida	Anencephaly	Spina bifida	
1979	21	10	3	1	35
1980	11	6	3	2	22
1981	13	9	4	2	28

Discussion

During the past three years the total number of amniocenteses in this region as well as the categories of indication appear to have become quite stable (table I). In 1979, the most recent year for which detailed national records are available,[1] there were 13 628 live and stillbirths in Lothians, Borders, and Fife. If it is assumed that about 3% of pregnancies miscarry between mid-term and delivery there would have been 14 050 mid-trimester maternities. The 690 amniocenteses performed in 1979 thus represent about 5% of the whole group, a percentage that does not appear to have substantially changed in 1980 and 1981.

A survey of antenatal diagnosis experience in the United Kingdom in 1976 showed that 1·1% of the pregnant population underwent amniocentesis.[2] It was calculated that complete availability of diagnostic services and full acceptance by high-risk mothers should lead to an amniocentesis rate of about 8%. This assumed that the proportion of mothers over the age of 34 would be above 7%. The Registrar General's Report for Scotland for 1979,[1] however, showed only 643 births in this region to this group of mothers, or 4·7% of the total. The 235 amniocenteses where the indication was maternal age thus represent a take-up of 37%. This low figure is surprising in terms of the availability of diagnostic services and the publicity given to the risks to the older mother.

In contrast, the acceptance of maternal serum α-fetoprotein screening appears to have been consistently good. During a limited pilot phase of screening from 1975 to 1977, 79% of mothers availed themselves of the service.[3] The service was transferred to a district hospital laboratory in 1978 and the catchment area widened to cover all mothers in Lothians, Borders, and Fife. The take-up was 67% in 1979 and over 80% in 1980 (S Stein, personal communication).

As shown in table II the indication leading to the highest proportion of terminations was raised maternal serum α-fetoprotein. This is not surprising since earlier studies have shown that a mother with two sequential α-fetoprotein values above the 95th percentile of the normal range has a 10-15% chance of carrying an abnormal fetus.[4] The remaining indications in table II all lead to about the expected number of terminations, though the category labelled "other" was somewhat swollen by six cases of anencephaly initially detected by ultrasonar scan.

Recently there have been several criticisms of the role of maternal serum α-fetoprotein screening in antenatal care.[5 6] One programme in East Anglia was discontinued after two years' experience in the belief that α-fetoprotein had poor predictive value in detecting spina bifida.[5] On more solid grounds, other investigators have pointed to the declining incidence of neural tube defects in the United Kingdom[7] and to the promising results of periconceptional vitamin supplementation in primary prevention.[8] Edwards[9] has even argued that a substantial reduction in incidence would make screening uneconomic. Though there seems little doubt that the incidence of neural tube defects has declined in recent years (whether temporarily or permanently is not clear; see table III), and though wide-scale vitamin supplementation may also eventually contribute to a further decline, the conclusion that α-fetoprotein screening should be suspended seems both premature and fundamentally misdirected.[9]

The mistake is to view maternal serum screening out of context. It should correctly be seen as a method of segregating out from a whole population those mothers who have an increased risk of bearing a child with a serious birth abnormality, and therefore be compared with other methods of identifying high-risk pregnancies, such as knowledge of maternal age or family history of children with birth defects. In this light the achievements of α-fetoprotein screening have been remarkable. In this area it has consistently reached and been accepted by more than three-quarters of pregnant women. In contrast screening of mothers aged 35 or more for Down's syndrome has reached only about one-third of the relevant group. Furthermore, α-fetoprotein screening can detect about 80% of neural tube defects, while complete acceptance of antenatal diagnosis by older mothers will probably locate only some 20% of cases of Down's syndrome.[10] It is therefore not surprising that 63% (66/104) of the terminations listed in table II resulted from α-fetoprotein screening. When the 12 terminations based on serum screening but where no amniocentesis was performed were added to table II, the proportion was 67% (78/116). Even if all cases of anencephaly are discounted (on the doubtful grounds that it is a lethal disorder) maternal serum α-fetoprotein screening was still responsible for most of the abnormal fetuses detected (35/63, 56%).

It is thus clear that a serum α-fetoprotein screening programme can make a major contribution to the reduction in birth incidence of children with serious abnormalities. None the less such programmes are not without problems. They require a considerable degree of organisation and maintenance, and good communications between laboratory and obstetrician. They may generate considerable anxieties among mothers whose serum α-fetoprotein values lie above the defined action point,[11] and thus demand expert forms of counselling and reassurance. The sensitivity of screening for spina bifida is only of the order of 80%,[3 12] and missed cases may elicit resentment among mothers who had hoped for protection against the birth of an affected child. Despite these imperfections, maternal serum α-fetoprotein screening remains the only proved method of reducing the risk of a mother delivering a child with a neural tube defect.

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Problem awaiting solution

I have encountered a problem recently which I feel should be brought to the attention of the DHSS. As a general adult psychiatrist I am actively concerned in treating patients with anorexia nervosa, many of these as outpatients. Apart from various therapies, drug and behavioural, it is, of course, a problem to be sure how much food these patients are actually getting down. A simple and rational way of controlling this is to use high-calorie carbohydrate supplements that are acceptable to the patients. One of these is Hycal, available in various flavoured solutions or, even better, Caloreen, which is a tasteless powder and can be sprinkled on food.

Unfortunately, these are borderline substances and can be prescribed only for certain defined conditions—namely, renal failure, liver cirrhosis, disaccharide intolerance, amino-acid disorders, malabsorption states, and proved hypoglycaemia. Plainly, anorexia nervosa does not fall into these categories and Caloreen and Hycal cannot, therefore, be supplied on prescription. It could be argued that the patient should be expected to buy these products, but such is the emotional importance of food in these cases that one can see that the patient may choose to use the non-availability of these high-calorie foods to reinforce her argument about why her weight is not increasing.

I have found that these high calorie supplements are a painless and easily monitored way of dealing with this illness on an outpatient basis, and anything that can reduce the need to subject the patient to the sort of inpatient struggles that all hospitals have experienced with anorexic patients must be a positive advantage in the treatment of anorexia nervosa. I would suggest that anorexia nervosa be added to the list of conditions for which Hycal and Caloreen can be prescribed.—M A LAUNER, consultant psychiatrist, Burnley.