although one who was not told until her child was over 4 years old made the comment that she might not have had another child if she had realized earlier that her first child was mentally defective. The proportion of mothers expressing satisfaction with the time they were told fell from over 80% of those told shortly after birth to 22% of those told after their child was 2 years old.

The mother's social status made little difference to the time when she was first told that her child was a mongol. Rather more of those in social classes I, II, and III were told before one month after birth, but by one year equal proportions in all social classes had been informed.

Mothers of firstborn infants were less likely to be told early than mothers of later-born infants. When the mother had other normal children one in three had been told before one month and only one in ten had not been told by the child's first birthday. When the mongol was a firstborn one in four of the mothers was told in the first month and one in three not until after the first year. Only one of 12 mothers of firstborn mongols had no criticisms to make about the time or method of telling compared with 32 out of 58 mothers of later-born mongols.

Nearly one-half of the total mothers said that they realized before being told that there was something seriously wrong with the child's development. This was no less common when the mongol was a firstborn child. Five mothers recognized the stigmata of mongolism when they first saw their babies; one suspected that something was wrong because "a lot of doctors" (most probably students) seemed interested in the baby, and three others because of their transfer to a single room or special kindness shown by nursing and medical staff. One mother read her case notes.

After discharge from maternity hospital five mothers experienced considerable difficulties in management and feeding but were given no explanation of the cause; two of these were mothers of firstborn mongols and had attributed the difficulties to their own inexperience. Nine mothers stated that they could have been spared months of uncertainty and unexpressed fears if they had been told earlier. Only three complained that they had been told in an abrupt, blunt, or unsympathetic manner, which makes it unlikely that many of the criticisms voiced stemmed from resentment felt against the individual who first revealed that the child was mentally defective.

To sum up, the mothers who spoke most appreciatively about the way they had been told were those who, having been warned or told soon after birth, were given a full explanation at that time or within the next three months, more especially if thereafter they were encouraged to return to the family doctor or paediatrician with any further queries or problems and were given regular support and advice throughout the early years. The mothers who were still most resentful were those who had suspected that their child was not normal, had sought advice on this account, and had been reassured that there was nothing seriously wrong.

Summary

During an aetiological study of severe mental subnormality in Edinburgh, mothers of mongol children supplied information about when and how they first learnt of their child's defect and their reactions to the time and method of telling. A full explanation given in the early months, coupled with regular support thereafter, appeared to facilitate the mother's acceptance of and adjustment to her child's handicap.

This investigation has been carried out with the support and co-operation of Dr. R. Short, Senior Medical Officer for Mental Health Services, Public Health Department, City of Edinburgh. We are indebted to him for his assistance and also to Dr. C. F. Drysdale, Assistant Medical Officer of Health, Public Health Department, City of Edinburgh; Dr. I. F. Craik, Chief Executive School Medical Officer for the City of Edinburgh; and his secretary, Miss A. E. M. Lewis. One of us (E. M. W.) was supported by a grant from the Mental Health Research Fund.

Medical Memoranda

Haemolytic Anaemia Associated with Ovarian Teratoma

Brit. med. J., 1964, 2, 1307–1308

Ovarian tumours are known to be associated occasionally with haemolytic anaemia, but the association is apparently a rare one, only nine cases being recorded. A further case is reported here.

Case Report

An unmarried woman, aged 48, was admitted to Aberdeen Royal Infirmary on 7 April 1963 with a three-month history of jaundice and increasing dyspnoea on effort. She had never been jaundiced previously, and there was no family history of jaundice.

She was well nourished, pale, and moderately icteric. The liver edge was 2 in. (5 cm.) below the costal margin and the spleen was easily palpable. A tumour was noted in the lower abdomen slightly to the left of the midline, and was found on bimanual examination to be a firm rounded cystic swelling about the size of a grape-fruit. Radiographs of the pelvis showed a dermoid tumour about 4 in. (10 cm.) in diameter that was more than half-filled with fat and contained several teeth.

Investigation showed a severe haemolytic anaemia. Prednisone in a dose of 60 mg. daily was given from 6 to 14 May, but the haemolysis was unaffected and on 14 May six units of packed cells were transfused.

On 23 May Dr. J. F. B. Wyper removed a large left-sided ovarian tumour. The right ovary appeared perfectly healthy, as did the uterus apart from the presence of a small fibroid. She made an excellent post-operative recovery. The haemoglobin level was well-maintained and three weeks after operation there was no evidence of excessive haemolysis. This was confirmed at repeated reviews during the following 12 months.

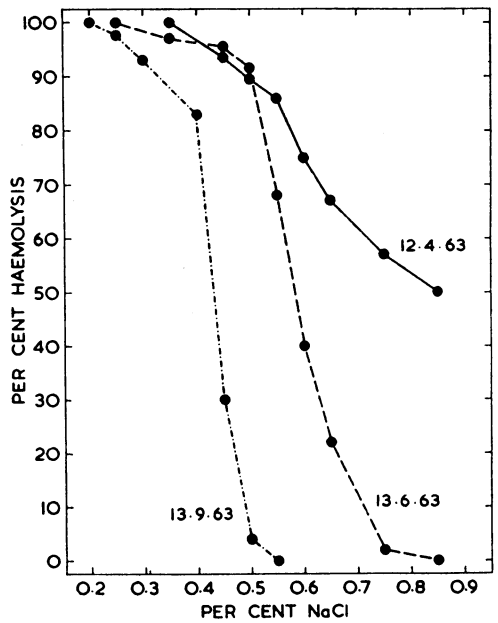
Her married sister, aged 59, was examined. She showed no evidence of excessive haemolysis, and the appearance and osmotic fragility of the red cells were normal.

Laboratory Investigations.—Haematological and biochemical data are presented in the Table. The osmotic fragility of the red cells on three occasions is shown in the Fig. The patient's blood was Group A Rh-positive. The direct Coombs test in serial dilutions was negative. No autoagglutinins were detected at 4° C., room temperature, or 37° C. No alpha-, beta-, or auto-haemolysins were detected at room temperature or at 37° C. The blood Wassermann reaction was negative. Examination of the sternal marrow showed very marked normoblastic and macronormoblastic hyperplasia. The

ovarian tumour was a benign multilocular cystic teratoma. It measured 12.5 × 11 × 9 cm. and contained teeth, hair, and sebaceous material. Microscopically the following tissues were noted: stratified squamous epithelium with sebaceous glands, thyroid tissue, fibrous tissue, and glandular tissue.

Haematological and Biochemical Data

	Haemoglobin g./100 ml.	Reticulocytes/ 100 R.B.C.	Spherocytosis	Leucocytes/ c.mm.	Platelets/ c.mm.	Serum Bilirubin mg./100 ml.	Faecal Urobilinogen mg./100 g. Faeces
1963:							
April 9 ..	4·2	97	++	10,950	92,000	5·1	
12 ..	4·6	97	++	12,200	115,000		
26 ..	4·8		++				
May 1 ..							946
3 ..	4·8		++	8,400			1,408
6 ..	*Started prednisone 60 mg. daily*						
7 ..	5·0	98	++	8,350	241,000		
11 ..	4·4	87	++	13,550	382,000		
13 ..	3·6	58	++	12,900	290,000		
14 ..	3·8	45	++				
14–15	*Transfusion. 6 units packed cells*						
17 ..	11·7	11	+	42,000	159,000		
22 ..	9·2	12	+		102,000		
23 ..	*Removal of ovarian teratoma*						
29 ..	11·9	5	+		325,000		
June 7 ..	12·3	6	+	10,400	406,000		166
13 ..							110
15 ..							74
July 10 ..	12·4	1·8	0	10,300			
Aug. 9 ..	12·4	< 1	0	4,350		0·2	
Sept. 12 ..	14·0	< 1	0				
1964:							
April 4 ..	13·6	< 1	0				



Osmotic fragility of red cells on admission and at three weeks and 16 weeks after removal of ovarian teratoma.

COMMENT

The immediate response of the haemolytic anaemia to the removal of the teratoma strongly suggests a causal relationship. The possibility that the patient was suffering from an exacerbation of haemolysis due to hereditary spherocytosis is excluded by the absence of a personal or family history of jaundice, by the complete and sustained remission after removal of the tumour despite retention of the spleen, and by the normal haematological findings in her sister.

Since the first description (West-Watson and Young, 1938), nine cases of haemolytic anaemia associated with ovarian tumours have been reported. The case described by Sandøe (1953) and by Ahrengot (1953), though it was included in a review by Müller and Schubothe (1958), is not considered to be a true example of this association, since the cyst was located in the mesentery and was too degenerate for histological diagnosis. In the case of Watson (1939) and perhaps also that of Lindeboom (1950) the causal relationship is somewhat doubtful. The ages of the remaining seven patients have varied from 4 to 54 years. In six cases the tumour was a benign teratoma or dermoid cyst (West-Watson and Young, 1938; Singer and Dameshek, 1941; Allibone and Collins, 1951; Wuhrmann, 1954; André *et al.*, 1955; Müller and Schubothe, 1958), and in one case (Jones and Tillman, 1945) it was a pseudomucinous cystadenocarcinoma. The haemolytic process is of gradual onset and pursues a chronic course, the anaemia often being profound at the time of presentation. The spleen may not be palpable and is rarely markedly enlarged. In the six patients in whom it has been done, the direct Coombs test was positive in two, weakly positive in one, and negative in two, one of these being the present case.

In all seven patients prompt cessation of the haemolysis followed removal of the ovarian tumour. Both cortisone and A.C.T.H. were without effect in two cases (Wuhrmann, 1954; Müller and Schubothe, 1958), and cortisone was used without success in the case of André *et al.* (1955).

The precise mechanism that provokes the haemolytic process in these cases remains obscure. There is no evidence to suggest that the cyst produces antibodies, none having been demonstrated in the tissues or contents of the cysts in the cases described by Allibone and Collins (1951), André *et al.* (1955), and Müller and Schubothe (1958). These last authors consider it more likely that the cyst is the source of antigens which stimulate the production of antibodies acting among other things against the erythrocytes. They suggest that such antigens could originate from the protein stagnating in the cyst, perhaps through the splitting of peptide links; unstable lipids and lipoproteins which are frequent constituents of the cyst fluid might also be antigenic. While the factors which determine the haemolytic potential of a particular teratoma are not clear the composition of its contents and the vascular arrangement may be important.

Though the association with haemolytic anaemia is rare, ovarian teratomata are relatively common, constituting 11% of all ovarian tumours (Baird, 1962). Since the prognosis in cases showing this association is so good the possibility of an otherwise symptomless ovarian tumour should be considered in a woman with unexplained haemolytic anaemia.

I am grateful to Dr. K. N. V. Palmer for permission to publish this case.

G. M. McANDREW, M.B., M.R.C.P.ED.
Department of Medicine,
University of Aberdeen.

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