and anal mucosae. She was edentulous at the first visit, and we were unable to find out if she had any dental abnormality, such as has been described by various authors (Gerth and Flegel, 1956). The salivary-gland enlargement, which has previously been described, was unlikely to be mumps, even though she had been a contact, as the clinical course was dissimilar.

Since 1943 endocranial calcification has been noted in some of these patients, and fits, thought to be associated with it, occurred in two cases. The calcification appears to be in the temporal lobes, even in Ramos e Silva's (1943) case. Our patient had bilateral calcification in the temporal lobes, probably medial to the posterior horns of the lateral ventricles; but it was not considered justifiable from the patient's point of view to confirm this by air studies. Her fits were diagnosed as akinetic epilepsy; there was no auditory or gustatory aura as might be expected in temporal-lobe lesions, and electroencephalograms were normal.

The marked bruising tendency with a positive tourniquet test has not previously been described in lipoid proteinosis. It was suppressed completely while she was taking corticosteroids. Probably her menorrhagia was an associated phenomenon. Full investigation of the purpura revealed only increased capillary fragility. This could be explained by the widespread deposition, in and around capillaries, of eosinophilic staining material, rather analogous to primary systematized amyloidosis.

The histological findings leave no doubt that our patient has the same disorder as that described by Laymon and Hill (1957) and by Ungar and Katzenellenbogen (1957), although the changes are less advanced than those described and illustrated in the latter paper. We cannot help to elucidate the nature of the hyaline material, which in its perivascular distribution bears a resemblance to amyloid but in its staining properties shows wide differences. The localization of lipid to the degenerate orcein-positive fibres appears significant, and, so far as they go, our findings seem to support those of Ungar and Katzenellenbogen.

Lipoid proteinosis has been regarded as a systemic disease because of the presence of eye changes (Holtz and Schulze, 1950; Gerth and Flegel, 1956; Scott and Findlay, 1960), association with diabetes mellitus (Urbach and Wiethe, 1929; Tompkins and Weinstein, 1954), testicular involvement (Bazex, 1939), intracranial calcifications and bromsulphthalein retention (Wile and Snow, 1941; Price *et al.*, 1947).

Diabetes mellitus was found only as an associated feature in early cases, and the testicular involvement (Bazex, 1939) took the form of deposition of sudanophilic material around blood-vessels. A degree of bromsulphthalein retention is insufficient by itself to enable liver involvement to be postulated; and liver biopsy of two cases showed normal histology (Tompkins and Weinstein, 1954). Extensive skeletal x-ray films of one case (Price *et al.*, 1947) were normal. Full investigation of several cases has not borne out this possibility.

A study of the literature indicates that, unlike primary systematized amyloidosis, the prognosis of lipoid proteinosis is good; the only potentially lethal complication being laryngeal obstruction. Corticosteroids have proved beneficial to skin and laryngeal lesions in our case and one of Eberhartinger and Niebauer's; early and prolonged administration may be of benefit.

Summary

The first British case of lipoid proteinosis of Urbach and Wiethe is described. In addition to the usual features the patient had an increased tendency to bruise, probably caused by infiltration of blood-vessel walls.

Histopathology and histochemistry support the hypothesis that the disease is primarily an infiltrative process.

Early and prolonged corticosteroid therapy may be of benefit.

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A STUDY OF 65 CASES OF COMPOUND PRESENTATION

BY

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Although a few isolated cases of compound presentation have been reported in recent years (Claye, 1929; Wong, 1932; Henderson, 1952; King and Mitchell, 1953; Bowles, 1954; Anderson, 1955; Alderman, 1958), only two studies of relatively large series of cases have been presented in the English language—namely, one by Goplerud and Eastman (1953) and the other by Quinlivan (1957). The former made a survey on 65 cases and the latter on 33. A study was therefore made of the 65 cases which occurred in the Tsan Yuk Hospital, Hong Kong, over a period of 14 years (1946 to 1959). Compound presentation is said to occur when one or more extremities, an arm, a leg, or both, prolapse alongside a head which presents, or when one or both upper limbs prolapse alongside a presenting breech.

Incidence

The incidence as shown in the German literature in the nineteenth century varied between 1:1,956 and 1:64 (Pernice, 1858). McCormick (1947) gave an incidence of 1:250, Gibberd (1951) one of 1:800, Goplerud and Eastman (1953) one of 1:652, and Quinlivan (1957) one of 1:360 in St. James' Hospital, London, and one of 1:500 in Hillingdon Hospital, Uxbridge, Middlesex.

In the years 1946 to 1959 inclusive there were 85,879 deliveries in the Tsan Yuk Hospital. In this same period 65 cases of compound presentation were encountered,

thus giving an incidence of 1:1,321, a much lower incidence than those previously reported. Some cases may have spontaneously corrected themselves and therefore been overlooked. The several varieties of compound presentation encountered in this series are compared with Goplerud's series in Table I.

TABLE I.—*Varities of Compound Presentation*

	No. of Cases in Present Series	No. of Cases in Goplerud's Series
Vertex	57 (8)	55 (13)
Vertex and hand (arm) ..	51 (8)	49 (11)
" hand (arm), and leg ..	3	4 (1)
" and foot ..	3	2 (1)
Breech and hand ..	7 (2)	4 (1)
Face ..	1	6 (1)
	65	65

In Tables I-III the figures in parentheses denote cases that were associated with prolapse of cord.

Aetiology

Multiparity, size of foetus, multiple pregnancy, placenta praevia, contracted pelvis, and hydramnios were found to be important predisposing factors. All of them are in fact conditions which may prevent complete filling and occlusion of the pelvic inlet by the presenting part. Intervention such as version, either external or internal, performed for any of a variety of reasons, may, if not completed successfully, end in a compound presentation. None of the cases in this series had had a version performed before the diagnosis of compound presentation was made.

Fifty-four (83%) of the patients were multiparae, among whom 11 had had five or more previous premature or full-time deliveries.

Twenty-seven (41.5%) of the babies were premature (1,000 to 2,350 g.). This rate was five times the prematurity rate (8.4%) in Tsan Yuk Hospital over the same period.

In this series 18 (27.7%) cases were associated with twin pregnancy; the hospital incidence was 1% (or 1:98.5). The second twin was, as expected, more commonly involved (16 out of the 18 cases). Three (4.6%) cases were associated with placenta praevia; the hospital incidence was 0.6% (528 cases in 85,879 deliveries). Contracted pelvis was found in 4 (6.1%) of the 65 cases; the hospital incidence was 2.1% (1,852 cases in 85,879 deliveries). Hydramnios was present in two cases (3%); the hospital incidence was 0.2% (175 cases in 85,879 deliveries).

Diagnosis

Compound presentation may be suspected when the foetal head remains high and sometimes deviated slightly from the midline or when there is early rupture of membranes, but a definite diagnosis can only be made by either a vaginal examination or a plain x-ray film of the abdomen. If a compound presentation is suspected vaginal examination should be performed as soon as possible after rupture of the membranes to confirm or refute the diagnosis and also to ascertain any prolapse of the umbilical cord. The diagnosis of compound presentation was made at full dilatation in 41 (63%) of the 65 cases. None was made before the onset of labour.

Complications

Prolapse of cord is commonly encountered in cases with compound presentation. This is not surprising, for the predisposing causes of the one are exactly the

same as those of the other—namely, those conditions which prevent engagement of the presenting part. The hospital incidence of prolapse of cord during the 14 years was 1 in 282.5 cases (0.36%). Ten cases of prolapse of cord were encountered in this series of 65 cases of compound presentation, giving an incidence of 1:6.5 cases (15.4%). Prolapse of cord was therefore encountered more than 40 times as often in compound presentation as would be ordinarily expected.

Uterine inertia has been said to be a common complication of compound presentation. This has not been borne out in the present series of cases. The longest first stage lasted 38½ hours. The first stage lasted more than 24 hours in only three cases, 12 to 24 hours in 11, and less than 12 hours in 46.

Post-partum haemorrhage was present in 3 (4.6%) of the 65 cases, in contrast to the hospital incidence of 1.2%. Two of these occurred in twin pregnancies. Retained placenta which necessitated manual removal was present also in three cases, giving an incidence of 4.6%, in contrast to the hospital incidence of 0.8%. The association of post-partum haemorrhage and retained placenta with compound presentation may be just incidental.

Prognosis

The hazards to the mother are said to be substantially augmented by the danger of a rupture of the uterus. Reviewing the three cases of uterine rupture in their series of 65 cases of compound presentation, Goplerud and Eastman (1953) came to the conclusion that the uterine ruptures were the result of reprehensible obstetric management rather than of the compound presentation itself. Not a single case of uterine rupture occurred in the present series. Only three cases of maternal morbidity were encountered: one was a case of typhoid, one a case of breast engorgement, and one a case of possible genital infection.

Foetal prognosis is known to be bad and foetal mortality high. These facts are confirmed by a study of the present series. The neonatal death rate and the stillbirth rate were 16.7% and 16.9% respectively, in contrast to the respective hospital rates of 1.6% and 2%. But if cases of prematurity and prolapse of cord and one case of anencephaly are excluded, the perinatal mortality that can be attributed to compound presentation *per se* is low: 1 stillbirth in 33 cases (3.3%, or 33 per 1,000), which agrees exactly with the figure given by Goplerud and Eastman (1953). Prolapse of cord, prematurity, prolapse of foot with or without prolapse of hand, twin pregnancy, and the type of management are found to affect the foetal prognosis.

The foetal mortality was 40% in cases with prolapse of cord in contrast to one of 24.6% in cases without prolapse of cord. Prolapse of cord was relatively more common in premature than in mature deliveries.

Prematurity *per se* was responsible for a large proportion of foetal deaths. Sixteen of the 20 foetal deaths (neonatal deaths and stillbirths) occurred in premature babies. Of the 27 premature babies, 11 survived, 8 died neonatally, and 8 were stillbirths, giving a foetal loss of 69.3%.

There is general agreement that foetal mortality is greater when a foot prolapses than when a hand presents. As shown in Table I, of the 57 cases of vertex presentation, 51 were associated with prolapse of hand and 6

with prolapse of foot. The foetal mortality was 25.5% when the hand alone prolapsed, but 66% when the foot prolapsed.

There were two neonatal deaths and two stillbirths among the 18 twin deliveries, giving a foetal loss of 22.2%. Of the four deaths, three occurred in premature babies. Twin pregnancy *per se* does not appear to worsen the prognosis. As a matter of fact, a premature twin stands a better chance of survival than a single premature foetus (30% in contrast to an overall foetal loss of 69.3% in the premature group).

In 23 of the 65 cases no intervention was necessary, and deliveries were spontaneous. Of the 65 cases, 20 were treated only by replacement of the prolapsed limb; all of these 20 patients then delivered spontaneously. Altogether 26 attempts at replacement of the prolapsed limb were made. Three of them were followed immediately by forceps delivery and the other three were unsuccessful. Of the three unsuccessful attempts one was followed an hour later by a successful attempt, and the other two subsequently had spontaneous deliveries. Seven cases were delivered by internal version and breech extraction. Six of the seven cases of breech presentation were delivered by breech extraction (the other one was associated with placenta praevia and was delivered by caesarean section). In all, five women were delivered by caesarean section (including the one just mentioned) and four by forceps (three of them preceded by replacement of the prolapsed limb). The indications for caesarean section were: placenta praevia in one case, contracted pelvis in two cases, prolapse of cord in one, and contracted pelvis together with prolapse of cord in one. The foetal loss in relation to various types of management is shown in Table II.

In order to find out the effect of the type of management on the foetal prognosis, the foetal mortality is

TABLE II.—Foetal Loss in Various Types of Management

	No Treatment	Replacement of Prolapsed Part	Version and Extraction	Breech Extraction	Caesarean Section	Forceps
No. of cases	23	20	7	6	5	4
Living	16 (1)	16 (1)	1	5 (1)	4 (2)	3 (1)
Neonatal death	2	3	2	1 (1)	1	0
Stillbirth	5 (1)	1	4 (2)	0	0	1
Total foetal loss	30%	20%	86%	16%	20%	25%

TABLE III.—Foetal Loss in Various Types of Management (Excluding Premature Cases)

	No Treatment	Replacement of Prolapsed Part	Version and Extraction	Breech Extraction	Caesarean Section	Forceps
No. of cases	14	11	4	3	4	2
Living	13	11 (1)	1	3	4 (2)	2
Neonatal death	0	0	1	0	0	0
Stillbirth	1 (1)	0	2 (1)	0	0	0
Total foetal loss	7%	0	75%	0	0	0

TABLE IV.—Foetal Loss in Various Types of Management (Excluding Cases with Prolapse of Cord, Placenta Praevia, and Congenital Abnormality)

	No Treatment	Replacement of Prolapsed Part	Version and Extraction	Breech Extraction	Caesarean Section	Forceps
No. of cases	21	18	4	4	2	2
Living	15	15	1	4	2	2
Neonatal death	2	2	1	0	0	0
Stillbirth	4	1	2	0	0	0
Total foetal loss	29%	17%	75%	0	0	0

analysed in Table III, excluding cases of prematurity, and in Table IV, excluding cases with prolapse of cord and with placenta praevia, and one case of anencephaly.

After consideration of all the available data, the following suggestions are made: (a) The best method of managing cases of compound presentation without complications, such as prolapse of cord and placenta praevia, is careful observation with no manipulation. (b) Replacement of hand gives equally good results. (c) Version and extraction has a very limited place in the management of compound presentation. It is the most dangerous form of management from the viewpoint of both mother and child. (d) Breech extraction is advisable for cases where the breech presents, the cervix is fully dilated, and complications such as pelvic contraction are excluded. (e) Caesarean section is indicated in cases of placenta praevia, in contracted pelvis, and also when prolapse of cord is detected before full dilatation of cervix. (f) In cases of compound presentation with prolapse of cord, the management is that of prolapsed cord—namely, caesarean section before full dilatation of the cervix and forceps or breech extraction after full dilatation. (g) The correct management of twin pregnancy and delivery and that of premature babies will further reduce the foetal loss.

Summary

Sixty-five cases of compound presentation that occurred in the Tsan Yuk Hospital over a period of 14 years (1946 to 1959) have been studied.

As there were 85,879 deliveries over this same period, the incidence of compound presentation was 1 in 1,321 deliveries.

Multiparity, small size of the foetus, multiple pregnancy, placenta praevia, contracted pelvis, and hydramnios were found to be important predisposing factors.

The diagnosis of compound presentation was made at full dilatation in almost two-thirds of the cases.

Prolapse of cord is commonly encountered in cases with compound presentation.

Compound presentation *per se* does not seem to affect the mother adversely. Foetal prognosis, too, does not seem to be affected by compound presentation, but it is adversely affected by prolapse of cord, prematurity, prolapse of foot, and the type of management given.

Some suggestions regarding treatment are made after consideration of all the available data.

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