symptom in Dercum's disease. Injection of pituitary gland extract and oral administration of thyroid gland extracts have given fair results in reducing the adiposity but neither relieves the pain. In most cases the patient has to be told to make the best of things.

Now Fröhlich's syndrome, dystrophia adiposo-genitalis, has been shown to be associated definitely with hypopituitarism, which may arise either from depression of the function of the gland by the pressure of a tumour in the neighbouring structures, or in another part of the pituitary itself, or from diminished activity. In view of the beneficial results that had been obtained by the x -ray treatment of acromegaly, it was natural that x rays should be given a trial also in Fröhlich's syndrome. The results obtained have been, as might be expected, very variable. In fact, Holfelder⁶ condemned x rays outright on the ground that the syndrome was well known to be due to a depression of function, which presumably x rays would further depress. Nevertheless good results have been obtained. Probably what happens is that in cases which do well the depression of function is associated with destruction or pressure by a tumour and the tumour is affected by the radiation, thus relieving the pressure. This is the view upheld by Baensh⁷ in a recent article in which he shows that whereas the cells of the adult pituitary are insensitive to radiation those of the growing animal and of adenomata are sensitive. Reviews of cases of Fröhlich's syndrome treated by irradiation of the pituitary are given by Kupferle and Szily, 1926,⁸ and by Marburg and Sgalitzer in their work *Die Röntgenbehandlung der Nervenkrankheiten*, published in 1930.⁹ They collected forty cases which showed variable results, but, on the whole, most showed some improvement in one respect or another. Adiposity was in general uninfluenced, genital function was improved, diabetes insipidus improved, headache was relieved, and eye symptoms, where present, were relieved in just over half the cases. Since then further papers have appeared by Schulte,¹⁰ ¹¹ Baensh,⁷ and Harris and Selinsky.¹² Recently cases of Cushing's syndrome have responded well to irradiation of the pituitary.¹³ ¹⁴

No case of Dercum's disease has, however, been found which has been treated by irradiation of the pituitary. Maddox¹⁵ says that he is going to try it, and that the method has not yet been fully exploited. A case of Cushing's syndrome—basophil adenoma—reported by Dr. R. D. Lawrence and treated with x -radiation by Dr. Hugh Davis is stated to have had "pains in the limbs" which disappeared after treatment. In view of the poor results of any therapeutic means hitherto tried, and of the strong probability of the pituitary being primarily at fault, it was suggested that radiation might be tried in this case. It was made quite clear to the patient and her husband that the treatment was experimental, but the patient was in such pain that she was ready "to try anything."

Treatment as follows was given at eight weekly intervals from February 8th to March 27th, 1934: 200 kV 4 mA, 6×8 cm., 0.8 mm. Cu + 1 mm. Al. Right and left temporal fields alternately. Four treatments to each. 140 r units each treatment.

The result was beyond expectations. The pain improved after the second treatment, and has now almost entirely disappeared. The adiposity has not been influenced, although subsequent dietary treatment has caused a steady reduction of weight. The patient says that she cannot stand long and gets readily faint and tired, and when she is tired her legs ache, but not like they did before. She thinks that the size of her legs has decreased. She had lost a little hair from the head, but this has grown again. With regard to the pain she says, "Life is worth living again; it wasn't before." The improvement

in the pain is maintained to the present date, eight months after treatment.

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CAESAREAN SECTION IN INFECTED CASES

A SERIES OF FORTY-FIVE CAESAREAN SECTIONS IN INFECTED OR POTENTIALLY INFECTED CASES, WITH NO MATERNAL MORTALITY

BY

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The risk of performing Caesarean section in infected or potentially infected cases is well known. Statistics quoted by Holland¹ give the maternal mortality as 9.4 per cent. where the operation is performed late in labour, 14 per cent. when it has been preceded by induction of labour, and 26.5 per cent. when it follows attempts at delivery by forceps or craniotomy. It is not, therefore, without a sense of serious responsibility that I admit having performed it in a considerable number of such cases. I report these, nevertheless, because indications are not lacking that the pendulum now tends to swing too far in the other direction, and that the lives of many infants and of a certain number of mothers are sacrificed, not because the operation is performed in potentially infected cases, but because it is not performed.

The possible dangers of a difficult instrumental delivery are not always realized, and those who refuse ever to perform Caesarean section in such cases as "failed forceps," for example, or the non-engagement of the head after a surgical induction of labour, may be unwittingly condemning their patients to a far greater evil.

It was the realization of this in my own practice, when, in a case which came under my care during labour, I had the misfortune as the result of a difficult delivery to lose both mother and child, that determined me to be less unwilling to perform Caesarean section in such cases in future. Apart from the maternal aspect of the case, the life of the foetus merits more consideration than it sometimes receives. It is difficult to understand why a life which is so jealously guarded, often at serious cost to the mother's health, during the first months of pregnancy should not be entitled to at least equal consideration during the last. To take only one illustration: it is well known that the foetal mortality in cases of prolonged uterine inertia, apart from organic obstruction, is very high. Bell,² in a series of forty-nine such cases occurring at Queen Charlotte's Hospital, reported a foetal mortality of 40 per cent., the maternal mortality in the same series being 10 per cent. And yet the condition of prolonged inertia is not one of the generally recognized indications for Caesarean section, the slight additional risk to the

mother being allowed to outweigh the enormously increased danger to the child. This consideration applies with special force to those cases where the pregnancy has either followed an operation for sterility, or the patient is an elderly primipara.

Case Records

The following details refer to a series of forty-five patients operated on during the past eleven years. I have endeavoured to trace every case which could come under this heading, but even if any have been overlooked I can state definitely that there has been no maternal death in any case which could have been included in this series.

The standard of puerperal morbidity is that described in *Queen Charlotte's Text Book of Obstetrics*,³ a puerperium being classified as morbid if the temperature reached 100.4° F. or more on two occasions between the second and eighth days.

The factors which have been taken as indicating potential or actual infection before the operation, and the actual cases concerned, are as follows.

I. Membranes Ruptured for More than Twenty-four Hours

Twelve cases. One foetal death (this foetus was believed to be dead before the operation). Four morbid puerperia.

This series includes two cases where the membranes had been ruptured for sixty hours or more, and a third where they had been ruptured fifty-two hours. (This patient had functional rigidity of the cervix and was very anxious for a child.) In a fourth case the membranes were known to have ruptured before the onset of labour, which had lasted four days, and were stated by the patient to have ruptured two weeks previously.

In many instances vaginal examinations varying from two to "about seven," or "several," had been made. Three of the cases were "trial labours," the membranes having been ruptured thirty-eight, forty-three, and "at least forty-eight" hours respectively.

The indication for operation was usually disproportion, but in one case, reported elsewhere,⁴ it was increasing toxæmia which failed to respond to treatment. Caesarean hysterectomy was performed once, the patient having been in labour six days, with the membranes ruptured sixty hours. On admission to hospital her pains were very strong, with practically no relaxation between pains, but the cervix was only two fingers dilated and the head was high. The child was believed to be dead, but Caesarean hysterectomy was considered safer than craniotomy.

II. Prolonged Labour Only

Five cases. No foetal mortality. One morbid puerperium.

One patient, who had had six vaginal examinations, had been in labour for five days, three for four days, and one, who had had three vaginal examinations, for thirty-eight hours. In two cases the delay was due to a rigid cervix, one patient being an elderly primipara—married for twelve years. In this last case a contraction ring was seen at the operation and divided before the child could be delivered. Inhalations of amyl nitrite had had no effect on it.

III. Surgical Induction of Labour

Thirteen cases. Two foetal deaths (one known to be dead before the operation). Six morbid puerperia.

In eight patients, three of whom had a morbid puerperium, labour had been induced by rupturing the membranes, and in five, of whom four had a morbid puerperium, by the introduction of bougies or a tube.

In nine cases the indication for induction was disproportion. In four it was severe toxæmia, but this did not respond to treatment, and it was feared eclampsia would supervene unless the patients were delivered immediately. In the case

where the child was known to be dead the head remained high and Caesarean section was chosen as being less dangerous than craniotomy.

IV. Ante-partum Haemorrhage with Preliminary Plugging or Douching

Five cases. One foetal death (macerated). Three morbid puerperia.

Four were cases of placenta praevia, the vagina having been plugged in three, and hot douching resorted to in one. The most interesting of these was a patient who was seized with sudden alarming haemorrhage in the ante-natal department after being examined. She became rapidly blanched and faint, and had to be plugged regardless of full antiseptic precautions. Despite a morbid puerperium she made an excellent recovery.

The fifth case had had vaginal plugging for profuse bleeding and when seen was almost moribund from loss of blood. Caesarean section was performed on the assumption that she was a case of placenta praevia, but without further examination for fear of causing more bleeding. She proved to be a case of severe toxæmia and accidental haemorrhage, the uterus being almost black in parts. The foetus was macerated.

V. "Failed Forceps"

Seven cases. One foetal death. Three morbid puerperia.

1. Multipara. Admitted from district, but forceps applied in hospital. Vagina plugged owing to bleeding following unsuccessful attempt at delivery. Non-morbid puerperium.

2. Primipara. Forceps delivery attempted in hospital after manual rotation of occipito-posterior position. Cervix and perineum torn. Child born alive, but died almost at once. Morbid puerperium.

3. Admitted to hospital as an emergency case with a history of "failed forceps" and many vaginal examinations. Cervix not fully dilated on admission, therefore further trial of labour given before operating. Morbid puerperium.

4. Primipara aged 42. Admitted to hospital as an emergency case, having had forceps applied eleven hours previously. Cervix not fully dilated on admission. Caesarean hysterectomy performed with drainage because the patient was very anxious for a living child. Non-morbid puerperium.

5. Primipara. Two attempts at forceps delivery in hospital, each preceded by attempt at manual rotation of occipito-posterior presentation. Cord tending to prolapse. Morbid puerperium. Lower segment operation.

6. Primipara aged 37. Anxious for a child. Seen in response to a message that attempts at forceps delivery had been continued for two hours. Unreduced posterior presentation. Vulva oedematous. Lower segment operation. Non-morbid puerperium.

7. Primipara. Anxious for a child. Labour complicated by fibroids. Forceps had been applied before the cervix was fully dilated. Lower segment operation and myomectomy. Non-morbid puerperium.

VI. Unclassified Cases

Three cases. No foetal death. Two morbid puerperia.

1. Prolonged labour with contracted pelvis. Had had "about ten vaginal examinations." Temperature 100° F. the night before operation. Non-morbid puerperium.

2. Second Caesarean section for contracted pelvis. Membranes had been ruptured for twelve hours, and a foot prolapsed outside the vulva before the operation. The foot was painted with violet-green before being replaced. Morbid puerperium.

3. Complicated by volvulus of caecum at term. Turbid fluid in peritoneal cavity. The volvulus could not be untwisted until Caesarean section had been performed (case reported elsewhere⁵).

Commentary

The total number of foetal deaths in the whole series was five, but of these two were known, and one believed, to have occurred before the operation. If these, with the three mothers, are excluded the foetal mortality of the

series is two out of forty-two, or 4.7 per cent. Nineteen cases—that is, 42.2 per cent.—had a morbid puerperium. In sixteen cases the lower segment operation was performed, and of these seven, or 43.7 per cent., had a morbid puerperium.

No attempt has been made to follow up the subsequent obstetric history of these cases as a whole, but I have performed a subsequent Caesarean section on three of the patients—namely, V 2, VI 3, and the patient with the rigid cervix referred to in I. It is interesting to note that the first of these became pregnant again after nine months, and the other two after five months, suggesting that the infection had not been a very serious one. In the case of V 2 there was an adhesion between the omentum and uterus, but none to the abdominal wall or elsewhere. In the other two cases there were numerous adhesions.

Technique of Operation

The technique adopted in both infected and clean cases is the same in many particulars. Thus in all my cases the abdomen is painted with Bonney's blue paint, and the vagina swabbed out when the patient is anaesthetized. Pitocin (1 c.cm.) is injected into the uterine wall immediately the child is delivered. If the classical operation is performed the uterus is sewn up with interrupted

silkworm-gut stitches, and a continuous catgut suture superimposed. The additional precautions taken in infected cases when the classical operation is employed include some or all of the following: evisceration of the uterus before incising it, swabbing its cavity with Bonney's blue paint, suturing an omental patch over the uterine scar, and abdominal drainage.

A prophylactic injection of scarlatinal antitoxin is given in every doubtful case, and if the soft parts have been injured—as, for example, in V 2—*B. welchii* serum is given too. Many cases receive quinine injections in addition. I now employ the lower segment operation in almost every doubtful case, but I have only done this comparatively recently, and if it is permissible to draw any conclusions from such a short series the results from the lower segment operation would not appear to be any better than those from the modified classical operation. When the risk of infection seems to be very great a Caesarean hysterectomy is performed, and this was done twice in this series.

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VON GIERKE'S DISEASE ASSOCIATED WITH AMYLORRHOEA

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A medical rarity has its own interest. If it can be shown to illustrate a perversion of a well-known physiological mechanism its value becomes even greater, in that it may shed light upon normal metabolic processes as yet imperfectly understood. In von Gierke's disease glycogen accumulates in various organs of the body, especially the liver, and is only broken down with the greatest difficulty.

The condition was first described by von Gierke¹ in 1929; subsequently cases have been recorded in America,^{2, 9} Australia,^{10, 11} France,^{12, 13} Germany,¹⁴⁻¹⁸ Holland,^{19, 20} Italy,²¹ and in this country by C. Worster-Drought,^{6, 7} Bellingham Smith and O'Flynn,⁵ E. C. Warner,⁴ and R. W. B. Ellis.^{2, 3} Von Gierke's¹ original case occurred in a little girl aged 8. It was characterized by progressive enlargement of the liver without any enlargement of the spleen. There was no jaundice and no ascites. The child died from influenzal pneumonia. At the necropsy the liver and kidneys were found to be greatly enlarged and infiltrated with enormous quantities of glycogen. The liver contained as much as 10 per cent. of its post-mortem weight of glycogen, and, as Schönheimer showed, this glycogen failed to undergo hydrolysis when it was incubated at body temperature. More recently cases have been described in which the deposition of glycogen has not been confined to the liver and kidneys, but has occurred in other tissues, such as skeletal,¹⁹ heart,^{8, 19} pyloric stomach muscle,²⁰ or even the central nervous system.⁸

The disease is one of childhood. It seems likely that there is a tendency to slow spontaneous recovery providing that the child does not succumb to an intercurrent infection. Stunted in growth, though by no means necessarily backward mentally, the child has a well-covered and prominent abdomen, which is in striking

contrast with its thin and underdeveloped limbs. There is progressive enlargement of the liver, but not of the spleen. There is no jaundice or ascites. The fasting blood sugar is low, and is not appreciably raised by the injection of adrenaline. The glucose tolerance curve is abnormal. Acetone is present in the urine at some stage of the disease, and in some cases is a constant feature.

Hypertrophic steatosis of the liver, which resembles the condition so closely that Debré and his co-workers¹² decline to distinguish between it and von Gierke's disease and group the two conditions together under the heading "hépatomégalies polycoriques," follows an acute infection, and does not give rise to ketonuria. Our object in recording this case is to describe a new finding and to discuss its bearing on the pathology of the condition.

Clinical and Biochemical Findings

The patient was 5 years old, and presented all the typical features of the condition except that, so far from being fat, he was definitely thinner than normal. The liver was greatly enlarged, especially the right lobe, which was separated from the left by a "notch" (see illustration). His weight had been average at birth, but had only increased erratically. Every cold caught was followed by a period of stationary weight lasting for six months or more. In January, 1934, following a minor infection, he had what was apparently an attack of ketosis, since he was comatose, with marked air-hunger. He was very bright mentally, but walked late, and until recently used to become extremely exhausted after short walks or other similar exertion.

Biochemically the usual features were present.

1. The fasting blood sugar was 50 and 57 mg. per cent. respectively on two occasions.

2. The glucose tolerance curve was abnormal in that it rose to a higher level than normal and took a long time to come down:

Fasting blood sugar	...	...	...	50 mg. per cent.
Forty-five minutes after the ingestion of 1 gram glucose per kg. of body weight	...	...	...	175 " " "
Two hours after glucose	...	...	...	100 " " "

3. There was a deficient response to the injection of adrenaline.

Fasting blood sugar	...	...	...	57 mg. per cent.
Fifteen minutes after the injection of 5 minims of 1 in 1,000 adrenaline,	...	...	...	
blood sugar	...	...	...	60 " " "