five weeks after exposure, and good fluorescence was observed throughout both feet, even in toes which were already blackened. As an indication of expected tissue loss it was useless, but it certainly revealed the presence of a brisk circulation and in so doing gave strong support to Lewis's theory of direct thermal injury.


### Summary

An account is given of two cases of trench-foot which occurred as a result of abnormal exposure in peacetime England.

It appears likely that trench-foot, immersion foot, and frostbite are but variants of the same clinical condition, the most important aetiological factor being in all cases a direct noxious effect of cold.

Recent experimental evidence favours early rapid warming to body temperature as the most effective form of treatment, and, provided suitable facilities are available to avoid overheating, the clinical application of this principle is advocated.


Our thanks are due to Dr. K. O. Black for permission to publish Case 1 and Dr. B. Branford Morgan for permission to publish Case 2. We are grateful to the Director of the Meteorological Office, Air Ministry, for details of weather conditions at the relevant times; and to Mr. N. K. Harrison for the photographs.


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The Institute of Dermatology held an open day on April 21 at St. John's Hospital for Diseases of the Skin, London. The object was to show visitors, both medical and lay, the research which is being done there. This includes research into the reaction of normal and abnormal skin to light which has made it possible to define light sensitivity in terms of very narrow wavelengths; work on the use of radioactive isotopes in the treatment of haemangiomata; investigations on epidermal sensitivity and the use of patch tests, particularly for cement and alkali dermatitis and cheilitis due to lipstick; and studies of the application of histochemical techniques to the detection of enzyme activity, and of cytodiagnosis to the diagnosis of skin lesions. There were also demonstrations of the L.E.-cell phenomenon, of methods of diagnosing scabies and ringworm of the nails, and of research on the penetration of ringworm fungi into hair keratin. Other studies included a survey of the serum proteins in skin diseases, particularly psoriasis, and an investigation of the changes in the nucleic acids and respiratory enzymes brought about by ultra-violet irradiation of the skin. There was also an exhibition of ointment bases. Although the main object of the open day was to demonstrate research, the opportunity was taken to display the very large collection of coloured photographs and wax moulages which is being built up for teaching purposes.

# THE POTTER SYNDROME OF RENAL AGENESIS


BY

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In 1946 Potter published a series of 20 cases in infants in whom bilateral absence of the kidneys was associated with a hypoplasia of the lungs and a characteristic facies. Her description of the facies, unfortunately labelled by some as the facies renalis, cannot be bettered, and reads: "The face most characteristically exhibits an increased space between the eyes, a prominent fold which arises at the inner canthus and sweeps downwards and laterally below the eyes, an unusual flattening of the nose, excessive recession of the chin, and moderate enlargement and decreased chondrification of the ears. The face gives a suggestion of premature senility." Thus, as in mongolism, recognition of this unusual appearance and knowledge of its implications are of the utmost importance in prognosis. Such children are generally regarded as rare occurrences, but recent experience in this unit suggests this may not be so, and details of five such cases are given in the accompanying Table. Two of the mothers are known to have had normal children subsequently.

### Discussion

These five infants demonstrate several features typical of this condition. Unlike most mothers of children with congenital abnormalities, all these mothers were primiparae and, at the ages of 20, 20, 21, 28, and 30, none remarkably late in child-bearing. In three, presentation was by breech, again quite frequent with this abnormality. In three a lack of amniotic fluid was apparent, and this, too, is exceedingly common, though not universal. Selby and Parmelee (1956) have described the case of an infant with this condition born tightly encased in intact membranes devoid of fluid, whilst in other reports the quantity of fluid appears normal, so that, whatever the origin of amniotic fluid, it can receive no more than a contribution from the foetal urine, as has been suggested in the past. There is a strong sex linkage, most instances being in males, and the child is most often born prematurely, weighing less even than the dates might lead one to expect. About two-thirds (Davidson and Ross, 1954) are liveborn but live for only a few hours, as the lungs are incapable of supporting life. Only a few have lived beyond a day, but Woolf and Allen (1953) have recorded an exceptional instance of a child living without kidneys for 39 days. Renal agenesis has been recorded several times in two members of the same family (Schmidt *et al.*, 1952; Baron, 1954) and in one of twins (Levin, 1952) —the twins shared the same placenta and were thought probably to be identical.

As might be expected, other congenital malformations are often found in these infants, but their pattern is again unusual, the majority involving the lower limbs and to a less extent the anus and external genitalia. Neither the typical facies nor the lung changes occur in every case, although most of those described since 1946 have done so. Of the 232 cases collected by Davidson and Ross (1954) only about 20% had no other notable evidence of abnormality and a further 10% had talipes, toe deformity, and other minor abnormality. Most of the lower-limb abnormalities are gross: absent kidneys are commonly associated with apus, monopus, and sirenomelia (the mermaid deformity). Whilst many of these are associated abnormalities, some may be secondary to the lack of amniotic fluid. Denis Browne (1955) believes abnormal mechanical intrauterine

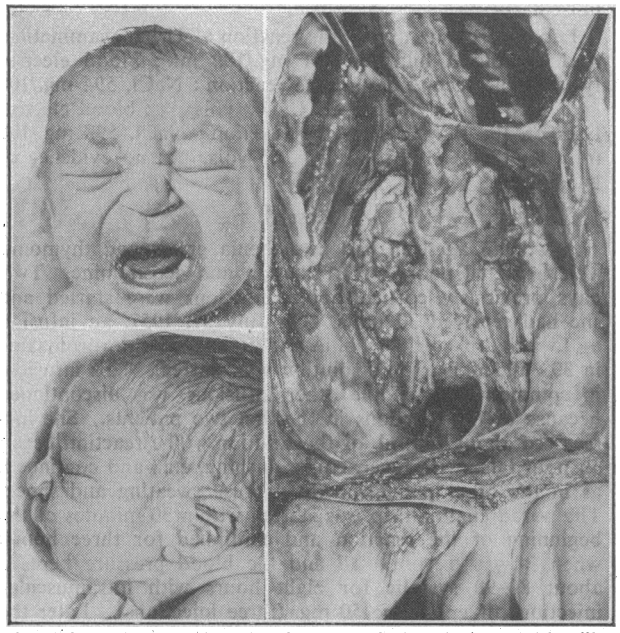
*Details of Cases*

| | Case 1 | Case 2 | Case 3 | Case 4 | Case 5 |
|---|---|---|---|---|---|
| Mother's age | 20 | 21 | 30 | 28 | 20 |
| Pregnancy | First | First | First | First | First |
| Blood group | O, Rh positive | A, Rh positive | O, Rh positive | O, Rh negative | O, Rh positive |
| Presentation | Breech | Vertex | Breech | Breech | Vertex |
| Delivery | 5 days after version to vertex at 34th week | Spontaneous at 33rd week | Caesarean section at term (foetal distress) | Spontaneous at term | Spontaneous at term |
| Quantity of liquor | Reduced | Reduced | " Very little " | Not remarkable | Not remarkable |
| Infant's weight | 3 lb. 10 oz. (1,640 g.) | 3 lb. (1,360 g.) | 4 lb. 13 oz. (2,180 g.) | 3 lb. (1,360 g.) | 4 lb. 4 oz. (1,927 g.) |
| ,, sex | Male | Male | Male | Male | Male |
| ,, condition | Stillborn (died during labour) | Blue asphyxia | Asphyxiated. Atelectasis | Stillborn (died during labour) | White asphyxia |
| ,, duration of life | — | 15 hours | 4½ days | — | 115 minutes |
| ,, facies | Typical | Typical | Typical | Normal | Typical |
| Necropsy: | | | | | |
| Renal tract and other pelvic organs | Kidneys, ureters, renal arteries absent. Minute bladder. Seminal vesicles not identified | Kidneys, ureters absent. Atretic bladder. | Left kidney, ureter, vas and seminal vesicle absent, right ureter and aplastic kidney present (see photograph). Right vas and seminal vesicle present. Small bladder without exit | Kidneys and ureters absent | Kidneys and ureters absent. Bladder not identified, rudimentary prostate |
| Lungs | Premature | Small and collapsed | Expanded, slight hypoplasia | No information | Airless and hypoplastic |
| Other findings | — | Abnormal tricuspid valve (two cusps only, thick, fused with short thick chordae) | — | — | Bilateral talipes. Flexion deformity metacarpal joints left hand. Cysts of brain |

With the exception of the mother of Case 5, who had a thrush vaginitis in early pregnancy and anaemia and a prolapsed disk later, there was nothing of note in the family histories, and their health before and during pregnancy was normal.

pressures cause such abnormalities as talipes, scoliosis, and other malpositions of the limbs, whilst an increase in hydrostatic pressure, by interfering with venous return, will produce stiffness, contractures, and perhaps even arthrogryposis, all found in these children. Malformations of the heart appear to be uncommonly associated with this condition. Davidson and Ross (1954) discovered reports of two examples of coarctation of the aorta (infantile type), two of double inferior vena cava, five of the various septal defects, one each of aortic atresia, pulmonary atresia, and transposition, and three of unspecified abnormality.

Both kidneys and ureters are usually absent, the position of the kidneys being occupied by flat disk-like adrenals. Where there is a ureter some identifiable renal tissue may develop, the stimulus of the ureteric bud from the Wolffian duct being apparently necessary (in humans) before differentiation of nephrogenic tissue into glomeruli and secretory tubules can occur (see photograph). Other Wolffian duct derivatives are usually present, as are the gonads. In the female the backward extension of the Müllerian ducts is often inhibited, resulting in absence of the uterus and upper vagina. The bladder is usually present as a narrow thin-walled sac extending up towards the umbilicus; the rectum and lower bowel or anus are not infrequently undeveloped or imperforate. The hypoplastic lungs resemble those of a younger foetus. It is generally considered that the cause of these abnormalities is the action of some blighting agent on the developing foetus at about the fourth to sixth week, and in this connexion both Osmond (1955) and Sylvester and Hughes (1954) have reported instances where the mother has confessed to taking abortifacients at this time.



Case 3. Photographs showing the facies and the disk-like adrenals, right ureter, and aplastic kidney.

In an attempt to discover the frequency of this condition the records of the last ten years were examined, and these five cases were discovered—the earliest in 1949. During these ten years there have been approximately 6,580 deliveries, with 166 neonatal deaths and 211 stillbirths (not excluding the occasional foetus of less than 28 weeks' gestation). In the same period there have been nine anencephalic children and nine with severe intestinal abnormality (obstruction, exomphalos, etc.), but it must be pointed out that this hospital, as a teaching hospital, accepts an unnatural proportion of primiparae and complicated pregnancies. Potter (1946) gives an incidence for the Chicago Lying-in Hospital of 0.3 per thousand births, whilst in this country Sylvester and Hughes (1954) found four examples in 9,440 deliveries, and Bound *et al.* (1956) 2 in 10,028 deliveries (Butler, personal communication).

## Summary

Five infants with bilateral renal agenesis have been born in the maternity wards of a teaching hospital in the last ten-year period, an incidence of 0.75 per thousand deliveries and 1.6% of the stillbirths and neonatal deaths. The case histories and post-mortem findings are summarized and a brief review of the salient features of this curious abnormality is given.

I am grateful to Dr. J. Forest Smith for permission to publish these case records and to Drs. H. Spencer and J. Macmillan for details of their findings at necropsy.

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