advantage—that is, to use enucleation operations only when there is a serious contraindication to the perurethral route.

Summary

Perurethral prostatic resection has been used in the treatment of over 90% of patients fit for the removal of prostatic obstruction.

We followed up 461 cases for periods of one to thirteen years, and in the great majority an excellent functional result has been maintained throughout.

Recurrent prostatic obstruction occurred in 6% of cases of benign hypertrophy followed for five to ten years and in 2% followed for one to five years.

Post-operative stricture occurred in 2% of cases and was not difficult to treat.

The operation failed to relieve incontinence in five cases and produced incontinence in two—one a nodular hyperplasia and the other a fibrosis of the prostate and periprostatic tissues of inflammatory origin.

The results suggest that the complications of perurethral prostatic resection have been exaggerated, and that this operation is worthy of a much larger place in British surgery.

Our debt to the surgeons of the Mayo Clinic is obvious. In addition we wish to thank Dr. A. B. M. Currie, Dr. W. Kraszewski, and the staff at the Victoria Infirmary, Glasgow, and Hairmyres Hospital, Lanarkshire.

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INCONTINENTIA PIGMENTI

REPORT OF CASE, WITH SUMMARY OF 29 CASES FROM THE LITERATURE

BY

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Twenty-six years ago Bloch (1926) demonstrated to the Swiss Dermatological Society at Basle an 18-months-old girl with irregularly distributed bizarre-shaped pigmentation of slate colour involving the trunk and lower extremities. After histological study of the skin he gave the condition the name of incontinentia pigmenti. In 1925 Bardach had reported "systematized naevus-formation" in a pair of 17-months-old twins, undoubtedly with the same condition. Since 1926 at least 29 other cases have been reported, the details of which are summarized below. Although reports have come from Germany, Sweden, the United States (both white and black subjects), Canada, Holland, Denmark, and Japan, no case has yet been recorded in England. It is true that Kalz and Mackenzie (1947) published a case in the *British Journal of Dermatology and Syphilis*, but the child was Polish and in Montreal.

The case reported here is that of a baby who was born and lives in Hertfordshire.

Case Report

A female infant, born on February 24, 1950, was the survivor of first-born twins, eight weeks premature, the other twin being stillborn. The mother and father are unrelated, are both healthy, and there is no family history of skin disorder. The mother's father and her younger brother were each blind in one eye from babyhood. Neither survives, and the nature of the blindness is unknown.

The patient weighed 3 lb. (1.4 kg.) at birth, but was rather feeble. I saw her in her first week for this reason, and she remained in hospital for five weeks. During this time her skin was normal. Except for the skin condition described below she has been a normal girl and free from symptoms. When aged 2 months the left lower limb became swollen and red, and when two or three days later the swelling subsided a "scorch-mark" was seen by the mother. At 7 months there was another attack, the child becoming ill-at-ease and feverish, and seeming to be in pain. The left lower limb became swollen, red, and shiny; after three hours blisters appeared in the pigmented areas and a measles-like rash on the clear skin. A few days later the swelling subsided and further pigmented areas were found. When I saw her at 13 months (March 28, 1951) there had been three attacks and the lower limbs only were affected. Three

FIG. 1.—Showing the pigmentation on the chest and the narrow stripes round the trunk.

FIG. 2.—Section of skin showing a large number of melanophores containing closely packed melanin granules. ($\times 240$.)

months later, after further attacks, there was reticulated dark-brown pigmentation on the chest and abdomen.

Being quite unfamiliar with the appearance, I referred her to my dermatological colleague at St. Bartholomew's Hospital, and Dr. Brian Russell suggested the correct diagnosis.

The child is making normal progress, though her weight is only 18 lb. (8.2 kg.) at 20 months. There are no abnormal pigments in the urine. Hair, teeth, nails, and eyes are normal. There is as yet no sign of fading.

Fig. 1 shows the curious pigmentation, with the particular concentration on the chest, characteristic of the condition. There are many parallel transverse narrow stripes round the trunk. Dr. Cureton has reported as follows on a biopsy specimen of skin taken from the left chest wall by Mr. R. S. Murley: "Sections of the skin show no gross structural changes. The amount of melanin pigment in the basal layer of the epithelium is variable—one section shows a moderate amount, another shows virtually none. All sections show a large number of melanophores containing closely packed melanin granules and situated almost entirely in the superficial corium. The appearances are those described in *incontinentia pigmenti*" (Fig. 2).

Summary of Cases

Twenty-nine case reports in the literature have been studied and are summarized, together with the thirtieth case, described above.

Sex.—Twenty-six are females, three are males, and the sex in one is not stated.

Condition of the Skin.—At birth:—18 were normal, 5 had erythema with or without blisters, 2 had actual pigmentation, and 2 others had a pigmented naevus. The condition in 3 is not stated. **In the neonatal period:**—Skin lesions were reported in 11 cases (erythema in 8, skin sepsis in 3). **Pigmentation** was present at birth in 2 cases, and developed from a skin lesion or *de novo* within the first month in 9 cases, before the age of 6 months in 6 more cases, and by the age of 3 years in a further 11. The 2 remaining cases were the father of two other cases and a girl first seen at 6½ years. **Colour** is variously described as steely grey or blue, slate-grey, dark brown, light brown, or café-au-lait. Several colours may be seen in one case. **Distribution:**—The upper part of the lower limbs and the trunk, particularly in the breast region, are most often and most heavily affected, and the lower part of the lower limbs and the upper limbs less often. Lesions of the scalp have twice been recorded. In two cases (Bardach, 1925; Siemens, 1929) a yellowish band of pigment was found on the face; and in one (Fleck, 1950) there were pigment patches on the soft palate. Sometimes the distribution is symmetrical; sometimes one side of the body is more marked than the other. **Fading** is reported in 12 cases, all but one being over the age of 3 years. No comment is made in 2. In 16 there was no fading, but 9 of these were under the age of 3 years at the date of the report.

Family History.—A family history of pigmentation occurs in Bardach's (1925) twins, who were both affected, and in Nägeli's (1927a, 1927b) father and two daughters. The present case is one of twins, the other being stillborn.

Associated Congenital Defects.—Nineteen cases have no other abnormality. Six have some congenital eye defect, such as pseudo-glioma or opacity of the lens. Other defects noted are brittle or spoon-shaped nails, patchy alopecia, dental hypoplasia, congenital heart disease, microcephaly, and spastic paraplegia. The record of congenital defects in other members of the family is certainly incomplete. Sulzberger *et al.* (1938) give a family tree showing congenital blindness in many females. In my case two males in the family had some form of congenital blindness.

Histology.—The most striking feature of the histological picture of the established condition is the collection of melanin pigment in the chromatophores of the corium, often arranged round the blood vessels. There is also a disarray of the basal cells of the epidermis. In scattered areas the regular line of these cells is broken, and individual cells are seen to be small or distended and vacuolated, with deficiency or absence of the normal pigment granules. The nuclei may be pyknotic. In the earlier stage of the disease there is some oedema of the skin, with vesiculation and perivascular cell infiltration (Haxthausen, 1944). In one post-mortem report (Seidlmayer, 1942) the adrenal medulla was aplastic.

Discussion

The explanation given by Bloch (1926) is that the melanin pigment in the skin, instead of leaving the basal layer and passing out into the stratum corneum to be flaked off in wear and tear, behaves "incontinently" and descends into the corium, where it is gathered up by the chromatophores and produces "autochthonous tattooing." There is no excess of melanin production. Bardach (1925), whose cases are the earliest known at present, regarded the condition as a form of systematized naevus. Nägeli (1927a, 1927b) used the name "familial chromatophore-naevus." Siemens (1929), who gives a very clear and careful account of the histology, regarded the basal layer lesion as the primary defect and suggested the title "melanosis corii degenerativa" as a description of what was seen in the skin section in preference to Bloch's name, which described a theory of production that might prove mistaken. He believed that this is an idiopathic example of degenerative melanosis which can occur under other circumstances, such as in parapsoriasis and poikiloderma.

Incontinentia pigmenti is a disorder of the skin arising at or soon after birth or in the early months of life, and usually in girls. It would be interesting to know how many of these subjects have been small or premature. Heilesen (1948) has stressed that there are three stages of the disorder, but that in some cases only the second stage may be present. First there is erythema with some papule or vesicle formation. Then when this subsides the affected area is noticed to be pigmented. Periodic attacks leave pigmentation more and more widely spread. In some cases the pigmentation appears in skin previously normal. The third stage—fading—may be incomplete or absent. At its height the pigmentation is found on the trunk and lower limbs in all cases. Upper limbs and scalp may be affected, as may the oral mucosa. Two cases had curious yellowish bands of pigment on the face. The colour is sometimes grey or steely blue or some shade of brown, several colours occurring in one case. The pattern is symmetrical, but often one side is more heavily involved than the other. All writers describe bizarre shapes, tree-like branching, and, round the subject's trunk, parallel lines giving a superficial and erroneous suggestion that nerves are followed.

A congenital defect of the eye or an ectodermal defect may be present in the case or in some member or members of the family. Other members of the family may have incontinentia pigmenti.

The familial character and the association with other recognized congenital ectodermal defects supports the view that this is a congenital abnormality of the skin.

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CONGENITAL DUODENAL OBSTRUCTION AND MONGOLISM

BY

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We have recently been impressed by the frequency with which intrinsic obstruction of the duodenum in the newborn has been associated with mongolism. During the past 25 years 32 infants with congenital atresia or severe stenosis of the duodenum have been admitted to Great Ormond Street, and of this number no fewer than 10 were recognized as mongols.

Case Reports

Case 1.—A first-born male child, three weeks premature. Mother aged 25. Admitted aged 6 weeks because of vomiting and constipation from birth. *Clinical diagnosis:* Mongolism and pyloric stenosis. *Operation:* Gastroenterostomy for apparent "duodenal ileus." Death occurred a few hours after operation. *Necropsy:* Mongolism; stenosis of third part of duodenum, peritonitis, and organized pleurisy.

Case 2.—A male child, tenth in family, born at seven months' gestation. Admitted aged 2½ days with imperforate anus, severe vomiting, and deep jaundice with purpura. *Clinical diagnosis:* Mongolism with anal atresia. X-ray film suggested possible duodenal atresia. *Operation:* Relief of anal atresia. Death occurred the following day. *Necropsy:* Mongolism; atresia at junction of second and third parts of duodenum; large subarachnoid haemorrhage.

Case 3.—A full-term male child, second in family. Admitted aged 1 week with vomiting. *Clinical diagnosis:* Mongolism with duodenal atresia. Died on tenth day. *Necropsy:* Mongolism; duodenal atresia just below ampulla; partial patency of foramen ovale.

Case 4.—A male child born six weeks before term after surgical induction for hydramnios. Eighth child; mother aged 36. Admitted aged 4 days with vomiting from birth.

Clinical diagnosis: Mongolism; prematurity and duodenal atresia. Death occurred on the sixth day. *Necropsy:* Mongolism; duodenal atresia immediately above ampulla; interventricular septal defect; cerebellum disproportionately small; undescended left testicle.

Case 5.—A male child, third in family. Admitted on day of birth with progressive jaundice. Rh incompatibility; direct Coombs's test positive. Typical mongol. *Clinical diagnosis:* Haemolytic disease of the newborn with mongolism. Death took place on second day. *Necropsy:* Mongolism; severe duodenal stenosis 1 cm. above the papilla; cerebral kernicterus; erythroblastosis foetalis; Meckel's diverticulum.

Case 6.—A first-born female child, two weeks premature; white asphyxia at birth following difficult forceps delivery. Mother aged 35. Admitted aged 3 days with vomiting from birth and passage of only mucus per rectum. *Clinical diagnosis:* Intestinal obstruction. *Operation:* Duodeno-jejunosomy for atresia at duodeno-jejunal flexure; lack of mesenteric attachment of ascending colon noted. Discharged home at 1 month of age after a stormy convalescence. Recognized as a typical mongol when seen in an out-patient clinic at 5 months of age.

Case 7.—A male child, third in family. Mother aged 36; suffered from hydramnios during pregnancy. Admitted aged 7 days because of persistent vomiting. *Clinical diagnosis:* Intestinal obstruction. *Operation:* Gastro-jejunosomy for duodenal atresia. Died shortly after operation. *Necropsy:* Mongolism; complete membranous occlusion of duodenum immediately distal to ampulla; extensive peritoneal haemorrhage; small cerebellum; undescended left testicle.

Case 8.—A female child five weeks premature, fifth of family. Mother aged 40; hydramnios during pregnancy. Admitted on third day with vomiting from birth. *Clinical diagnosis:* Atresia of duodenum. *Operation:* Duodeno-jejunosomy for atresia of proximal 4 in. (10 cm.) of jejunum. Died shortly afterwards. *Necropsy:* Mongolism; duodenal atresia immediately distal to papilla; multiple atresias of proximal jejunum; small intestine much shorter than normal; universal mesentery; cerebellum disproportionately small; partial patency of foramen ovale; failure of lobation of left lung.

Case 9.—A first-born full-term female child. Mother aged 25; hydramnios during pregnancy. Infant suffered asphyxia at birth. Admitted aged 5 days with vomiting, cyanotic attacks, and purpura. *Clinical diagnosis:* Duodenal atresia and possible mongolism. *Operation:* Peritoneal bands divided. Death on eighth day with oedema and cardiac failure. *Necropsy:* Extensive peritoneal haemorrhage; duodenal atresia immediately above the papilla; mongolism, small cerebellum; large auricular septal defect (septum secundum); pulmonary oedema.

Case 10.—A male child, eighth in family. Maternal age 39; hydramnios during pregnancy. Following severe birth asphyxia the infant began to regurgitate brown fluid. Admitted aged 5 days. *Clinical diagnosis:* Mongolism and duodenal atresia. *Operation:* Gastro-jejunosomy. Deterioration after tenth day with diarrhoea; death at 3 weeks of age. *Necropsy:* Mongolism; severe duodenal stenosis immediately proximal to the ampulla; universal mesentery; small brain; aberrant right subclavian artery; umbilical sepsis; oral thrush.

Discussion

Textbooks of surgery and paediatrics do not in general refer to this association, which in our experience has occurred with remarkable frequency, nor is it mentioned in the monographs of Benda (1947) and Engler (1949) on mongolism. In a survey of published cases of duodenal obstruction in the newborn Forshall (1947) found only five instances in which associated mongolism had been observed. More recently, however, Lanman (1949), in Boston, reported that, of 17 children with duodenal