



Pachyonychia Congenita Project

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We hope that making available the relevant information on Pachyonychia Congenita will be a means of furthering research to find effective therapies and a cure for PC.

of bronchitis; his right radial pulse was rather weaker than the left; he slept badly at night.

On December 7th Mr. Heath ligatured the third part of the subclavian artery, and then tied the carotid artery above the omo-hyoid muscle. Both vessels were tied with silk. Each wound was stitched, drained with a small tube, and dressed with iodoform powder and salicylic wool. The right arm was wrapped up in cotton wool, and supported by a sling.

The temperature rose to 100.6° on the fourth day after the operation: it then fell, and was normal on the seventh day. The wounds were dressed on the second day, and the tubes removed. Both healed rapidly without any suppuration. For the first five days after the operation the patient was troubled with cough and difficulty in breathing; he had a good deal of bronchial secretion, which greatly hampered his breathing; his pulse was irregular and intermittent. On the sixth day the patient's condition was much better; his breathing was much easier, and his cough had ceased; his pulse, though still rapid (100 per minute), was regular in force and frequency. He got up for the first time on the twenty-second day after the operation. On the fortieth day the condition of the aneurysm was noted as follows:—The swelling above and internal to the right sterno-clavicular joint is much less than before the operation; the pulsation at these places is also less. The swelling extends more along the clavicle and the impulse over the middle of that bone is more marked. The aneurysm has decreased in size in its former position, but in all probability it has extended deeper and spread towards the arch of the aorta. On the forty-third day the patient's cough was more troublesome and interfered with sleep; he expectorated a good deal of watery mucus; his pulse was again irregular, and every third beat was missed. The patient was discharged on the fifty-first day after the operation. The condition of the aneurysm was unchanged since the fortieth day; there was still no radial pulse on the right side, but the arm maintained its proper temperature. The patient expressed himself as feeling fairly well; he was better able to swallow, and his breathing was very much easier than it had been for several months before the operation; he did not require to be propped up in bed, as was the case before the operation; and he could move his right arm more freely than before.

KENSINGTON INFIRMARY.

A CASE OF PILEGMASIA DOLENS; NECROPSY.

(Under the care of Mr. VAN BUREN.)

Mrs. E.W.—, aged thirty-five, was admitted on March 5th. She had given birth to her ninth child three weeks previously. The labour had been long and tedious, and accompanied by severe flooding.

On admission the woman's condition was of a very grave character. She was markedly anæmic, and had that dusky heavy look so typical in severe puerperal disturbances. The right leg was considerably swollen from foot to hip; it was of a sallow colour, hard, not yielding to pressure, and the prominent superficial veins gave it a marbled aspect. The temperature was 104.5°; pulse 140; respiration 40. The urine was acid; sp. gr. 1020; no albumen. She complained of slight pain in the leg. The tongue was dry and fissured, and in a general typhoid condition. Sickness was a marked symptom, as was also diarrhoea. She stated that the attack had been ushered in, with shivering, headache, and general febrile sequelæ. She had never suckled the child, there being no secretion of milk from the first. The lungs and heart yielded no physical signs of disease. The spleen was thought to be enlarged, and there was slight tenderness on the right side of the uterus. The limb was ordered to be fomented with warm lead lotion and wrapped in cotton wool. The rest of the body to be sponged with iced water. Sulphate of quinine was administered in ten-grain doses every four hours, with a view of bringing down the temperature. Dover's powder was also given, in ten-grain doses twice a day, to control the diarrhoea and produce sleep. She was obviously in such a low condition that stimulants were certainly called for, and six ounces of brandy were ordered to be taken every twenty-four hours. The rest of the diet consisted of beef-tea, milk, eggs, and soda-water, which were ordered to be given iced. She kept the quinine down, and after sponging the tempera-

ture fell to 102°. The night nurse reported a fair night's rest, and cessation of sickness and diarrhoea. The temperature fell still further to 101.4°. She was certainly better when seen on the morning of the 7th. Pulse 128; respiration 32. On the evening of this day she had a rigor, which lasted an hour, and the temperature went up to 104.2°. Pulse 144; respiration 42. Diarrhoea set in, and sickness prevented food being administered by the mouth. Fine crepitations were detected at the bases of both lungs, but there was no cough or expectoration. Owing to pains in the leg, which the patient thought were aggravated by fomentations, these were omitted, and cotton wool only applied. She had little sleep that night. On the morning of the 8th she was in a sinking condition; diarrhoea and sickness still persisted. The nurse stated that there was a discharge from the vagina for the first time, which resembled ordinary menstruation. The pulse was very fluttering and feeble, and, gradually passing into a comatose condition, she died at 12 o'clock, the temperature just before death being 101.4°.

Necropsy.—The body was well nourished, the only external sign of disease being the swelling of the right leg. The bases of both lungs were in a state of engorgement, but there was no consolidation. The heart was normal, and no clots were found in the pulmonary arteries. The liver was distinctly fatty, as were also both kidneys. The spleen was slightly enlarged and in a state of active hyperæmia. On section, it was soft, diffident, of a dark-red colour, and extremely vascular. The uterus was found to be the seat of interesting changes. Distinct signs of endometritis were found in the mucous membrane. At the placental site, which was situated to the right, purulent infiltration of the uterine wall had occurred, and there had evidently been active inflammation at this site. On making a section pus welled out, and the surrounding tissues were considerably softened. In the right iliac and femoral veins thrombi were found.

Remarks by Mr. VAN BUREN.—Fatal cases of phlegmasia dolens are so rarely met with, and the opportunities of performing a necropsy necessarily so few, that when one does meet with a case that can throw any light on the pathology of the disease it should be availed of to its utmost limits. In the Kensington Infirmary during the last three years I have watched about 450 labours. Puerperal septicæmia has occasionally, unfortunately, been present, but it has never assumed the form of the present case. I look upon the placental site as the primary source of infection in this case. The history of flooding at the labour is important, as this would favour the tendency to thrombosis by interfering with the nutrition of the walls of the vessels. The placenta being situated to the right side explains the right leg being affected, and is in itself interesting. Had there been no swelling of the leg, I should have been unable to distinguish this case from one of ordinary puerperal septicæmia. The rigors, high temperature, and condition of the spleen point to this. The patient was addicted to alcohol, I believe, which would account for the appearance of the liver and kidneys; fatty degeneration of the latter organs is undoubtedly a rare occurrence. The inflammation of the lungs was probably the result of hypostatic congestion. I regret now that I did not syringe out the uterus with an antiseptic, and I shall certainly bear this in mind in future cases; and probably the loss of blood was so great at the confinement that transfusion would have been of service, and lessened the tendency to thrombosis.

LINCOLN COUNTY HOSPITAL.

CONGENITAL DEFORMITY OF THE NAILS (ONYCHOGRYPHOSIS).

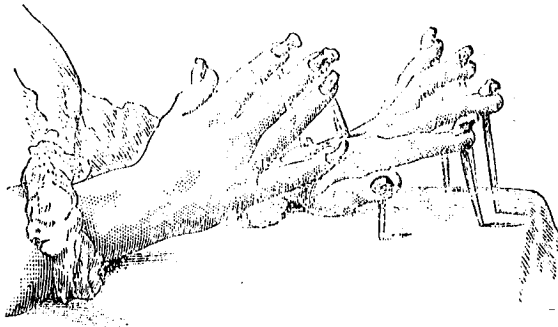
(Under the care of Mr. SYMPSON.)

In November, 1887, a school-girl (Susannah S.—), aged eleven, applied, on account of some slight ailment, as a casual patient at the Lincoln County hospital, when Mr. Matthew Johnston, the then house surgeon, noticing that she was the subject of a peculiar condition of the nails, referred her to Mr. Sympton.

On examination, the girl's hands were seen to be normal as to form and size. The nails projected upwards and forwards from their matrices to the distance of from one-half to three-quarters of an inch, so as to present the appearance of claws. The mother stated that the nails grew rapidly, and that when they were cut a quantity of clear fluid exuded,

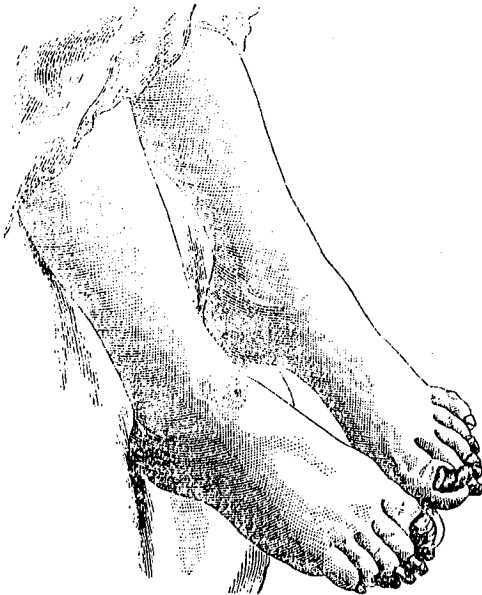
and they became very sore. (Fig. 1.) The feet were well formed, but the nails of the toes presented features similar to those of the fingers, excepting that they were shorter, being about a quarter of an inch in length. There were some abrasions and corns on the feet and toes, through

FIG. 1.



difficulty in obtaining boots that would fit. (Fig. 2.) There was some diffused redness with thickening and cracking of the skin over each olecranon. There were a few warts over the right supra-spinatus muscle. The mother said that the child had been subject to skin eruptions ever since she had been vaccinated. The child had not had cleft

FIG. 2.



palate; she cut her teeth, which were normal, in good time. She was either extremely reserved or else not very intelligent. The girl was born at full time. The malformation of the nails was present at birth. Her father and mother are healthy. She has five brothers and one sister, all of whom are healthy and well formed, and there is no evidence of her other relatives having any malformation whatever. The knee-jerk was slightly exaggerated on the left side, normal on the right. There was no ankle-clonus or supinator reflex. The electric reactions of muscle and skin were normal. Ophthalmoscopic examination revealed nothing abnormal, save that the choroidal ring was very distinct.

ILKLEY HOSPITAL AND CONVALESCENT HOME.—The twenty-sixth annual report for 1887 states that the number of patients admitted during the year was 968. No death had occurred. The working expenses were £1378 2s. 3d., and the total receipts £1032 12s. 3d., showing a deficit of £345 10s., which had been met out of income from other sources, and which had absorbed all the donations with the exception of two life donations. A resolution, thanking Dr. Johnstone, the medical officer, for his services, and that a gratuity of £50 be given him in acknowledgment of the same, was agreed to.

Medical Societies.

ROYAL MEDICAL & CHIRURGICAL SOCIETY.

Double Nephro-lithotomy.—Splenectomy.

An ordinary meeting of this Society was held on Tuesday last, Sir E. H. Sieveking, F.R.S., President, in the chair.

Mr. HERBERT W. PAGE read a case of Double Nephro-lithotomy, in which Lateral and Median Lithotomy had been previously performed, with remarks on Sympathy between the Kidneys. A man aged twenty-two entered St. Mary's Hospital in November, 1886. For ten years he had suffered pain in the region of the left kidney, with occasional attacks of left renal colic. In 1882 he began to pass gravel and to suffer from irritability of bladder, and in 1885 a small calculus was removed from the bladder by lateral lithotomy. Symptoms of vesical calculus soon recurred, together with pain in the left loin, and his general health became much broken. His urine contained pus and occasionally blood. Left calculous pyelo-nephritis was diagnosed in addition to stone in the bladder, and this was accordingly removed by median incision on Dec. 4th, 1886. On the 15th the left kidney was exposed in the loin and two small calculi were removed from a large suppurating cyst. The amount of pus, however, did not materially lessen, and towards the end of January he began to have pain in the region of the right kidney, from which it was soon suspected that the pus now came. On Feb. 18th he was attacked with violent pain in the right loin, with rigors, vomiting, and high temperature. At the same time the pus almost entirely disappeared, and his urine dropped from forty-two to twenty ounces. It was believed that this urine was furnished by the left kidney only. The symptoms increased and there was fulness in the right loin. On Feb. 25th the right kidney was exposed; it was found much enlarged and surrounded by fetid pus, and there was an opening into the pelvis from which an abscess had probably burst into the perinephral tissue. No stones were found. He did well after this operation. The urine steadily increased in quantity, the pus diminished, and his pain ceased. He left the hospital on May 26th, and a few days afterwards he passed four small calculi per urethram. From that time he made steady progress towards recovery. The case is of interest from its bearing on the question of sympathy between the kidneys, and it was suggested that in this case the right kidney had in all probability contained calculi, though without giving rise to symptoms, for some considerable time, and that its condition was revealed by the operation on its fellow. In support of this view the author referred to a case recently under his care in which the presence of calculi was first made known after the operation of hysterectomy, undertaken by the late Dr. Meadows. Reference was made to other cases in which there was a somewhat similar sequence of events. From another point of view Mr. Page considered this case of importance in its bearing on nephrectomy and preliminary abdominal exploration. Nephrectomy, as time proved, would have been in this case a very wrong thing, but he had never felt quite satisfied that abdominal exploration, by which he would have found an apparently useless left kidney and a right kidney so enlarged as to be presumably fit for all future needs, its calculi being, moreover, so small as to elude detection, might not there and then have led to nephrectomy. The patient would have been placed thereby in a position of infinitely greater danger when at a future day the right kidney was in its turn *hors de combat*. The case thus demonstrates with how many precautions and with what consideration abdominal exploration for the examination of kidneys must be made.—Mr. RICKMAN J. GODLEE concurred in the views expressed by the author, and urged the advisability of not performing nephrectomy. Disturbances in the abdomen, however, resulting, might mechanically explain the development of symptoms of calculus latent hitherto.—Mr. PAGE, in reply, considered there was no such thing as pure sympathy between the kidneys any more than existed between the kidney and any other viscus. In his case abdominal exploration might have led astray by finding the left kidney to be so diseased as to be useless, and even some of his colleagues advised nephrectomy, which would have been bad for the patient, since at the nephro-lithotomy on the right organ the diseased left kidney probably helped to carry off some of the