

4. The frequent formation of stricture either cicatricial or cancerous following operation necessitating inguinal colostomy.

5. The straightening and tension of the sigmoid destroyed it as a fecal container.

6. That sentiment and not chance has proven the main reason for continuing to place an uncontrollable anus in a comparatively inaccessible situation.

The gain in the combined operation has been in a selection of the operation to the case, either radical removal *en masse*, with all glands, fat and connective tissue or a colostomy for palliation. The retention of the sigmoid as a fecal container and the peculiar formation of the anus, giving a fair degree of control in an accessible situation.

SOME REMARKS ON SCLEREMA NEONATORUM,

WITH REPORT OF A CASE WITH AUTOPSY.

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Sclerema neonatorum is a rare disease. Up to the present time I have been able to find but seven cases reported in America, and a careful review of the literature of Europe affords but comparatively few examples of the strange condition. The obscurity of the etiology of the malady and the scarcity of clinical reports of cases with autopsies, as well as the meager and unsatisfactory descriptions of the same found in most text-books, render doubly interesting the report of a typical case, and afford an excuse for a perhaps too lengthy description of the clinical picture of the disease.

Uzembizius, a physician of Ulm, in a work entitled an "Ephemeris of Natural Curiosities," published nearly two centuries ago, described an infant, born at term, which from top to toe became rigid and cold, resembling a piece of flesh dried by fire. This is regarded as the first recorded case of infantile sclerema, and it was at that time ascribed to maternal impressions, since the mother of the baby had spent much time in the churches, worshipping the images and statues found there.

To Underwood, however, an English physician, belongs the credit of having first recognized and described the disease, a description, too, so vivid and so true as to have left little to be added or to be taken away. His observations were particularly noted in France, and the condition most thoroughly studied there, although for many years it was confounded with a very similar condition, edema neonatorum. This disease presents many features in common with sclerema, but differs in so many respects as to leave little doubt that they are essentially separate and distinct. Andry (1781), Leger (1825), Billard (1839), Villax (1849), all recognized, studied and described the malady and developed many and ingenious theories to account for it. It remained for Parrot, however, the brilliant Parisian pediatricist, to establish the fact that sclerema neonatorum was a distinct clinical entity, although the conclusions of this observer have been somewhat modified by the studies of more recent years.

The subjects of this disease are usually found among the premature, weakened, poorly-developed infants of the foundling asylums. Cases occurring in private practice are exceptionally rare, although Parrot cites cases developing in well-nourished, robust and well-fed infants. Whether or not hereditary syphilis plays a rôle in the etiology is still a disputed point. Cases have been

reported in which improvement or even recovery has followed treatment by mercurial inunction, but there is room for much skepticism as to the correct diagnosis in such instances. Within the past decade attempts have been made to isolate specific bacteria and to demonstrate that they were responsible for the development of the disease, but the number of observations in this direction is as yet too few for any definite conclusions to be deduced.

The affection, which is characterized by an induration of the skin and subcutaneous tissues, does not, as a rule, develop immediately after birth, but the symptoms are first noticed within the following seven to ten days, although according to Hennig they may be delayed as late as the seventh month. It is doubtful, however, whether such late developing cases should be regarded as instances of genuine sclerema neonatorum, but that they should rather be classed with scleroderma. In the first place, they do not correspond clinically with sclerema, they more frequently end in recovery and show a tendency for the induration to occur in isolated areas. It seems reasonable, therefore, to consider those cases which develop late as scleroderma infantum corresponding to scleroderma adultum, than to regard them as instances of true sclerema neonatorum as described by Underwood.

The symptoms may develop suddenly with no particular warning, may follow or complicate an acute disease, such as pneumonia, or they may be preceded by a variety of indefinite signs of general disturbance, such as vomiting, diarrhea with greenish stools, obstipation, convulsions, etc. The infant may become thin and emaciated, the eyes sinking into the orbits, the skin of the face taking on a sallow tint, becoming wrinkled, wizened, like the face of a little old man; or the disease may appear so suddenly that when first seen the characteristic induration may be the first and foremost symptom to which our attention is directed.

With such an onset the changes in the skin, which represent the essential feature of the disease, develop. In the invaded region the skin becomes swollen, shiny, losing its normal folds, appearing as if under tension, very like edema, but harder, firmer, showing little or no evidence of pitting on pressure. The elasticity and suppleness of the skin are gone; it is no longer possible to pinch it up into folds by the fingers; it is cold, clammy, greasy to the feel, for all the world like the outside rind of a ham. It is hard, as if glued down to the subcutaneous tissues; it is impossible to slide skin over subcutaneous parts, skin, muscle and underlying bone appearing to be fused completely together.

The color of the invaded areas varies from a dirty yellowish jaundice-like color to a livid blue black, the tints being comparable to the array of colors afforded by an area of ecchymosis slowly absorbed. The temperature of the part is always much reduced.

The amount of surface of skin involved varies within wide limits in different cases. It may remain circumscribed, death occurring to effectually check the progress of the disease. Again the skin of the lower extremities and trunk may be alone affected, or the whole surface of the body may take part in the process. The part which first shows the thickening may also vary. Most often the skin of the buttocks or thighs appears to be earliest diseased, from there spreading upwards and downwards on the trunk and on the lower extremities. It may then become general over the whole surface of the body. On the other hand, the skin of the face may be the first point

to show the induration, more particularly in the region of the cheeks.

As a consequence of the thickening and hardening of the skin and subcutaneous tissues, the affected parts become stiff and inflexible. If the face be involved, the mouth and cheeks are set as if moulded, mask-like, resembling the face of a doll, crying, sucking, swallowing being alike impossible. If the limbs be attacked the members are swollen, blue black to dirty brownish yellow in color, hard and rigid, extended or slightly flexed, but stiff and immovable. When the disease becomes general the infant lies motionless, with or without very feeble cry, as stiff as and still as death, with scarcely any respiratory movement and with an ever-weakening pulse. At this stage of the disease Trousseau and Parrot were able to lift the body by catching hold at either end, as one would raise a stick of wood, and it has been possible to balance the body on the radial surface of the hand placed under the back, just as one would balance a figure of marble or of metal.

Such, in short, are the characteristic symptoms of sclerema neonatorum. Variations in intensity and in extent of the induration occur, but correspond in general to the above description. In striking contrast, however, to this is the absence of general manifestations of disease of any of the vital organs. No heart or lung disease, no disease of the nervous system or gastrointestinal tract has ever been found that might be considered constant in or in any way causative of sclerema. There seems rather to be a universal prostration of all the vital functions. The malady runs its course slowly but fatally without giving the slightest clue in regard to the primary cause of the trouble. In the skin itself where the morbid process is mainly localized, there occur no microscopic changes that can be regarded as peculiar to or characteristic of the malady. Atelectasis of the lungs, congenital defects of the heart and morbid processes in the digestive tract have been described, but must be regarded rather as accidental complications than as having anything to do with causing the disease. Money described two cases complicated with paralysis, and concludes that the disease may be a trophic disturbance. Numerous hypotheses have been advanced to explain the peculiar clinical changes, but we must admit that from a practical standpoint we know little or nothing of their etiology, being able neither to retard nor to arrest its advance. Recovery rarely if ever occurs, the infant after illness of a few days to a week or more dying from the extreme prostration or asthenia.

Turning now from a general survey of sclerema, the history of a particular case may prove interesting.

J. K., full term infant of healthy parents, male, seven days old, at birth was quite cyanosed; extremities, ears, lips, face all were blue and livid, presenting the characteristic appearance of a "blue baby." Careful auscultation of cardiac area, however, discovered no audible murmur. Condition lasted forty-eight hours and gradually disappeared, leaving no trace. Baby nursed, bowels moved and urine voided in an apparently normal manner.

During night of seventh day baby cried almost incessantly and with no apparent cause, bowels moving and urine being evacuated as usual.

EXAMINATION.—The following morning the following facts were disclosed: The baby refused to nurse and seemed generally tender to the touch. Penis swollen and infiltrated with hard edema, so much so as to effectually close preputial opening. Skin of same was glossy, shiny and did not pit on pressure. Skin of scrotum was also swollen, hard and board-like, shiny and cold. Questioning revealed the fact that the little patient had not urinated since the night before. Percussion over hypogastrium revealed dulness and with a view to relieving

the bladder a hasty circumcision was done and an unsuccessful attempt made to pass a catheter. Under a hot bath and warm fomentations, however, the bladder relieved itself, but nevertheless the hard, board-like infiltration continued and was rapidly spreading. The cord had not yet separated from the umbilicus, but there was no evidence of infection or inflammation in its neighborhood. Temperature per rectum 98.5 F.

On the following day the hard induration was still present and had spread upward on the trunk and downward on the legs. The least touch seemed very painful, causing the baby to cry for some time, whereas otherwise he was quiet. Bowels moved sluggishly and small in amount, greenish in color; urine also small in amount, stained diaper light yellow. It was impossible to obtain any for examination. Temperature 97. At this time a blood examination showed 6,000,000 erythrocytes per c.mm., with no deformity of same and no nucleated reds. Owing to difficulty in obtaining blood it was impossible to count white cells; but from study of ordinary smears they appeared to be less than normal in numbers and mostly of mononuclear type. Cultures taken from the blood were negative.

The next day the sclerema had spread over the whole of the extremities and midway up the trunk. Skin was hard and leathery, could not be pinched up into folds, was densely adherent to subjacent, all seeming to be fused indefinitely together.

The following day, the fourth of the illness, the conditions were much worse. The infiltrated areas were turned a deep bluish black, particularly the dependent parts, and were painful on least pressure. Bowels moved and urine voided, but very scanty in amount. Child refused to take any but slight amount of nourishment by a spoon. Legs were flexed at about an angle of forty-five degrees, were stiff and firm, impossible to straighten or bend. Sclerema had extended on chest, but arms and face were not involved. Temperature 96, rectum. Death the same afternoon.

AUTOPSY.—This was made the following day at 9 a. m., fourteen hours after death. It is important, especially on account of the negative results. The following changes were noted. Body slightly emaciated. Postmortem rigidity not well marked. Surface of body presents a mottled bluish-black to yellowish tint, the former color being most developed over dependent parts and on skin of extremities. Skin of legs, thighs, feet, scrotum, penis, pelvic regions, extending behind up to middle of back and in front midway to umbilicus, is swollen hard, board-like, stiff, glossy, not pitting on pressure. Skin can not be pinched up into folds by the fingers, neither will skin glide over subjacent tissues. The dark blue-black color is most developed in region of anus and penis, the circumcision wound of which appears gangrenous. Umbilical cord separated and stump is normal looking with no signs of inflammatory reaction about it.

On section, subcutaneous fat exceedingly small in amount. Skin of lower abdomen distinctly thickened, being three times the thickness of that of upper abdomen. On section it appeared very firm and fibrous, of mottled orange to lemon yellow to grayish color, subcutaneous fat showing a distinct reddish tinge. No serum exuded from cut surface. Heart and lungs normal with no sign of any congenital defect. Liver mahogany color, extending four cm. below free border of ribs with no macroscopic change. Gall bladder distended with thick tenacious black bile, which could not be squeezed into duodenum, although dissection of common duct disclosed no obstruction. Stomach distended with gas extending below to umbilicus and 1½ cm. to right of median line. Urinary tract normal; spleen normal. Mucous membrane of small and large intestine pink, soft and velvety with no signs of swollen follicles or of ulceration.

Such in short, were the results of the postmortem examination. Cultures were taken from the heart; spleen and peritoneum, but there occurred but a few colonies of *Staphylococcus albus* and variety of the color bacillus. That the presence of these organisms was accidental and not due to general infection I feel convinced, because of

the lack of any symptoms pointing to an infective process such as fever, with enlarged spleen. Microscopic examination of tissues revealed no change. In the skin, however, corresponding to the marked clinical symptoms, we should expect marked pathologic peculiarities. No change, however, has been found at all commensurate with the macroscopic appearance. Northrup reported a typical case in which the microscope revealed nothing especially abnormal. Parrot found hardening and atrophy of the skin and rete malpighii, the cells appearing indistinct and forming a fused mass. The subcutaneous fat appeared atrophied and the interstitial fibrous connective tissues increased. The blood vessels were shrunken. In the present case the corium seemed thickened, the cells and fibers composing it having a homogeneous granular, hazy appearance, suggestive of cloudy swelling. The subcutaneous fat appeared normal, the interstitial fibrous tissue being little if at all increased.

Ballantyne found a "dried-out skin," some thickening of the layers with some diminution of the subcutaneous fat. The pathology and the etiology are very obscure. What relation these slight skin changes bear to the profound prostration of the disease is difficult to explain. Hensch examined the nervous system from two cases and found nothing abnormal.

From the date of its first description many and ingenious theories have been advanced to explain its occurrence. Underwood believed it was due to a spasm of the skin in consequence of breathing bad air. Leger thought it was due to an improper and incomplete development of the intestinal canal. Parrot taught that sclerema was not a disease, but only a symptom, an end-stage of a general condition of malnutrition, which he termed "arthrepsie," and that the thickening and drying up of the skin was due to the loss of fluids in consequence of diarrhea and to the absence of any reparative effort on the part of the other cells of the body. Langer, in 1881, advanced the theory that in consequence of the lowering of the body temperature, the palmitin and stearin, which form the principal part of the adipose tissue of infants, solidify, thus causing the thickening and hardness. This may be considered but a modification of the theory of Parrot that the reduction of temperature is consequent to the general condition of mal-nutrition and mal-assimilation, and following this heat loss solidification of the subcutaneous fat occurs. In other words, the skin changes depend purely on a chemical and physical change in the cutaneous fat, regardless of any vital activity on the part of the body cells. Were this true we might reasonably expect a like change to occur post-mortem in the case of infants. Ballantyne asserts that the skin changes are the result of the "drying out" of the skin incident to the diarrhea, and this view seems to be favored by Wiederhofer and Soltmann. This case, however, as well as that of Northrup and others, seems to prove that a preceding diarrhea is not essential to the development of the disease, nor a necessary symptom of its progress. Schmitt, in 1893, reported three cases in which he isolated micro-organisms, mainly staphylococci, from the tissues, and he maintained that sclerema is due to a general infection with a definite micro-organism, but this observation has not as yet been corroborated by other observers. Pathologic evidence, on the other hand, shows no change at all comparable to the changes observed in general infection with any known organism. It may be, however, that the immature tissues of the newborn react to bacteria and their toxins differently than the mature tissue of adults with which we are more ac-

quainted. It seems to the writer more plausible to regard the condition as the result of an auto-intoxication from improper functional activity of the gastrointestinal tract, the changes in the skin being secondary to the retention of the poison, and its direct action on the skin, incident to an attempt at elimination.

HYPERTROPHY OF THE SYNOVIAL FRINGES OF THE KNEE JOINT,

WITH A REPORT OF TWELVE CASES.

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This disease has been described from time to time by various writers in medical journals, but as yet there has been only slight mention of the subject in works on surgery. The different authors have included this condition in their description of floating cartilage, internal derangement of the knee-joint and chronic synovitis, but have given little consideration to it independently. In designating the disease the profession have used various terms, such as rheumatic joint, torn cartilage, strained ligaments and synovitis; and undoubtedly some cases of it have been diagnosed as tuberculosis of the joint, which it sometimes resembles in its first stages and for which it might easily be mistaken.

From the cases which have come under my observation during the past year and a half I have selected twelve, typical in character, the treatment of which has extended over a sufficient length of time to allow definite conclusions to be drawn regarding them. Eight of these cases I operated on; the remaining four were treated by mechanical means. The unsatisfactory nature of the information contained in the text-books has led me to endeavor to formulate my own experience and its results.

ETIOLOGY.

The etiology is obscure. In a few of the cases traumatism plays an important part, while in others it has been impossible to associate injury with the beginning of the disease. In order to explain this condition in those cases where it is not traceable to injury, the complicated movements of the normal knee and the intricate distribution of the synovial membrane should be considered.

The knee-joint, like all diarthrodial joints, is lined with a synovial membrane, except where the articular cartilages are in contact. Here no membrane is found, except at the edge of the cartilage, which the membrane overlaps for two or three millimeters before merging into the cartilaginous structure. The inner surface presents a smooth shining appearance, interrupted (especially where the membrane folds to pass from one surface to another) by the synovial fringes, which are villous structures of varying size and length, somewhat resembling intestinal villi, the largest being perhaps one centimeter long. They are richly supplied with blood vessels, each villus containing a convoluted twig of an artery. Some of the fringes are, however, merely hernia-like protrusions into the joint of small masses of fat covered by membrane, which fill up unoccupied spaces.

The articulation between each condyle and the opposed almost flat surface of the tibia does not exactly form a hinge-joint, though strongly resembling one; and extension and flexion, the movements of which it is capable, are produced by a combination of gliding, rolling and rotation. In the movement of extension the condyles move parallel to one another, both gliding and roll-