CHAPTER VIII.

GENERAL DISEASES OF THE SECRETIONS.

CACHEXIÆ.

(1.) Rachitis, Rickets, English Disease, Double Limbs.—By rickets is understood a developmental disease of the skeleton, in which a diminution of the calcareous constituents of the bones is the principal symptom. The earliest definite descriptions of rickets date from the middle of the seventeenth century, and were given by the English physicians *Whistler*, *Boot*, and *Glisson*. About this time reports of a new disease were heard from various parts of England, and a commission, consisting of the physicians just named, was appointed to investigate it thoroughly.

Since that time but little has been added to our knowledge of the pathology, or causes, or varieties, of rachitis, till some fifteen years ago *Elsässer* discovered the rachitis of the skull. The pathological anatomy has been considerably enriched and elucidated since then by the researches of *Kölliker*, *Virchow*, and *Hermann Meyer*.

Pathological Anatomy.—For the purpose of correctly comprehending the rachitic alterations, it is necessary briefly to recapitulate the physiological growth of the bone. Every tubular bone grows in length and thickness. It grows in length by new layers of cartilage-cells which constantly form between the epiphyseal cartilage and the bone, in which calcareous salts are then deposited. It grows in thickness by the addition of new layers of bony substances immediately beneath the periosteum, from the tissue by which the latter is cemented to the bone. As the growth in thickness is much more insignificant, and progresses slower than that in length, the disturbances of the physiological growth at the cartilaginous ends are also more striking and liable to occur.

While the bone is enlarging externally in every direction, by the addition of new elementary tissue, the medullary space within it also increases in circumference. Thus we have a constant new formation of bone externally, an absorption of bone internally. The femur of a child may with ease be put into the medullary canal of the same bone of an adult, so that, by the time the child has grown up, the original infantile bone has been completely reformed.

The physiological growth of a bone consists, then, in—

(1.) New structural cell-elements deposited on its upper surface.
(2.) Their prompt ossification; and in
(3.) Absorption taking place in the centre of the bone.

Rachitis consists in the suspension, or in the imperfect performance of the second function or process, while the first and third remain normal, by which various very striking and peculiar alterations in color, form, and consistence, become perceptible.

In regard to *color*, the rachitic bone is particularly distinguished by a dark-red color, which, on the skull, may even assume a bluish redness. The more livid the bone, the greater, as a rule, has been the duration and the degree of the rachitic disease. All the bones of the same skeleton are not always reddened in an equal degree, some are darker, others again are brighter in color, and from this alone it is readily seen that rachitis is no simple chemical process, but is due to a complicated anatomo-physiological condition.

No rachitic bone retains its normal *form*. All the sharp angles of the bone become rounded off, the tubular bones in all cases become shortened, they cease to grow in length, the epiphyses swell and become bulbous, a condition which is most plainly seen on the sternal ends of the ribs, which are curved in various directions. On the tubular bones, for example, on the ribs, simple curvings occur, but very frequently actual fractures, or, more correctly speaking, contortions of the bones occur, especially in those of the lower extremities. In advanced rachitis the external layers of the bones, as we will show more in detail in the delineation of osseous derangements, contain so little calcareous salts, that they cannot be completely broken.

The internal parts of the bone lying next to the medullary canal, formed before the appearance of the rickets, may, it is true, break, and do indeed very frequently break, owing to their attenuation, in consequence of the absorption that goes on within. The external portions of the bones, however, yield, and, though they bend, still do not break, and therefore no displacement of fractured ends can take place. The bones that are bent, after the manner of a quill or willow-twig, subsequently heal with a blunt angle. This bending of rachitic bones, and the subsequent angular deformity, may result from the action of the flexor muscles and from the superincumbent weight of the body.

The apex of the angle thus formed in the forearm looks outward and forward, that of the arm almost straight outward, that of the thigh forward and outward, and that of the tibia, which usually bends near the ankle-joint, straight forward.

When such an infraction is sawn through longitudinally, after complete recovery, compact substance will be found on the convex surface only, and on the concave a broad layer of spongy substance. The medullary canal is completely closed at the point of fracture, by thick bony extuberations, which subsequently become attenuated,

though they never disappear entirely. We shall speak more minutely of the alterations of form of the individual parts of the skeleton when we come to treat of symptomatology.

The diminution of the *consistence* of rachitic bones is very remarkable. Incisions may be made several lines in *depth*, and, when the disease is much advanced, the bones may even be cut through entirely without any very great exertion, and without notching the knife. These are the coarser anatomo-pathological signs of a rachitic bone.

When the affected skeleton is subjected to a closer examination, the following alterations, more or less marked on all the bones, will be found: the periosteum is thicker than usual, of a milky opacity in many places, and of a rose-red color. On attempting to pull it off, small and sometimes large fragments of bone will remain adherent to it; the bone is always dark red, and has a particularly rough external surface. This state of the periosteum is most distinctly seen on the frontal bones in craniotabes. The skull, in this case, is sawn, or may even be cut through with the knife, with the greatest ease; and on its posterior parts it is impossible to use the saw, for the spots, that have become attenuated to the thickness of a card, will yield, become depressed, and irregularly torn by the saw. From the section through the frontal bones, small drops of bloody serum exude; from the section of the temporal and parietal bones there will be less, and from that of the occipital there will not be the least of such serum. The frontal bones are always slightly thickened, sometimes to twice their normal thickness, and the anterior portions of the parietal bones in contact with the coronal suture partake in this thickening; while the posterior portions, on the other hand, are quite as often attenuated as in the normal state. Toward the lambdoidal suture, both it and the occipital bone become membranous in spots, which are of a yellowish-red color. The other parts of the bone which are not entirely wasted become extremely thin, of a bright color, and totally devoid of diploë. By holding the cranium up toward the light, the extent as well as the degree of this rachitic thinning, the craniotabes, is clearly seen.

If the calvarium is examined on its inner surface, numerous depressions are found, entirely on the occipital portion, answering to the *impressiones digitatæ*, each one of which corresponds to a cerebral convolution, whose pressure produced the attenuation of the bone, an atrophy indeed of the osseous substance. Finally, the *dura mater* and *pericranium* are in contact with each other, by which, in the dried preparation, the osseous tunics simply appear to be pierced and the membranes left intact. In these membranes, which resemble the

dried fontanel, some white opaque points are still occasionally to be seen, which, on close examination, prove to be masses of unabsorbed calcareous mater. *Elsässer*, in his treatise on "The Soft Occiput," delineates a calvarium with nearly thirty apertures. Such a specimen, however, must be looked upon as one of the most extreme instances. The pericranium is, where it is stretched over the apertures, as well as in their vicinity, opaque and hypertrophied (Pl. VI., Fig. 4).

The pathological history of the soft occiput is: (1), one of deficient deposit of the usual phosphates in the external osseous layers of the entire bony skull; and (2), of absorption of those portions of the bone which have been softened by the pressure of the weight of the brain.

On the epiphyses of the tubular bones, additional characteristic signs may be observed. When a longitudinal incision of the articular head of a long bone—the femur, for example—is made, a thicker layer of cartilage is seen than in the normal condition (Pl. VI., Figs. 1–3a), and the line between the bone and cartilage, instead of being straight, is very irregularly indented and undulating (Pl. VI., Figs. 1–3b). The apices of the undulations which jut out from the bone into the cartilage are intensely injected, and contrast strongly with the bluish cartilage. The microscopical and chemical examination of the broad, bluish transition-layers, between the bone and cartilage, proves conclusively that it is a bone which has been retarded in its ossification, in which no bone-corpuscles at all, and but few traces of calcareous deposits in particular, are found.

On the diaphysis of the tubular bones equally marked alterations take place. The periosteum is materially thickened, and cannot be pulled off smoothly from the bone. Some fragments of porous bone are always torn off with it, and adhere to its inner surface. Immediately beneath the periosteum, broad whitish or reddish layers are found, which present a fine porous, pumice-stone-like structure.

The trabeculæ of this mass, according to *Virchow*, stand like perpendicular radiæ upon the surfaces of the bone. Deeper still, these radiæ are seen to be interrupted, first by a white and dense line of cortical layer, which is parallel with the upper surface of the bone. Then follows a new stratum of the same material, of a reddish color, and with stronger radiæ, which are again intersected by a compact parallel layer. Thus these layers alternate with each other a variable number of times, but the radiæ of the spongious layers constantly grow thicker, the nearer they approach the medullary canal, and their interstices become larger and redder, while the parallel layers become denser and firmer.

The rachitic tubular bone is softest and most porous directly be-

neath the periosteum, and constantly grows firmer toward the centre. The hypertrophy of the periosteum, and the softened condition of the external layers, explain also the singular process of infractions, and the impossibility of detecting actual displacement of the fragments and crepitation. These are the most important statements concerning the pathological anatomy of these bones. They are exhaustively and thoroughly depicted by *Virchow* in his Archives, vol. v.

The chemical examination of rachitic bones has always shown a marked diminution of the phosphates and carbonates of lime; the calcareous salts, instead of constituting two-thirds, often only forming one-fifth of the dried bone. In the urine, on the contrary, the phosphates are found augmented from three to five fold.

This increase of the phosphates in the urine, and its diminution in the bones, are not to be regarded as a process in which the calcareous salts already deposited in the bones are redissolved, and then excreted by the kidneys. The salts once deposited in the bones remain in them; a small quantity only may, as a result of the absorption that occurs in the parts in the immediate vicinity of the medullary canal, again come into circulation. The new enlargements in the longitudinal and transverse diameter of the bones, however, do not receive any more calcareous salts, and the salts of lime introduced with nutriments find no consumption in the organism, but are immediately excreted by the urine.

Why the deposit of calcareous salts in the bones ceases is still enveloped in complete obscurity. It is certain, however, that it is not a simple chemical redissolving of the already-perfect bone by an acid, for otherwise its structures would, both on the periphery and in the centre, be alike deprived of calcareous salts, which is certainly far from being the case. The layers immediately adjacent to the medullary canals are much more compact and richer in salts than those of the periphery.

If the skeleton of a child who has recovered from rachitis be examined, the bones will still be found curved in various degrees, the skull large, its sinciput hypertrophied, and the individual bones are remarkably heavy. All the soft, spongy, bony masses that have formed during the rickets have become converted into dense, compact osseous structure, and this subsequent ossification exceeds in hardness even the normal bone, on account of which they have also been called sclerosis, and in extreme cases even eburneatio.

No constant alterations are found in the rest of the organs, but the lungs, in all cases of marked rickets of the thorax, exhibit acquired atelectasis, and severe bronchitis, already spoken of in detail in the chapter on pulmonary affections, page 298. The muscles are pale and

flabby, and in various places, especially in the heart, reveal fatty degeneration. The liver often displays a decided augmentation of fat.

Symptoms.—Rachitis is a tolerably acute affection, and generally appears on the head first, and always before the close of the first year of life. Next in frequency it is seen in the ribs, noticeable several weeks after commencing rachitis of the skull, and, lastly, in the lower extremities, the pelvis, and spinal column.

Formerly an especial prodromatory stage was assumed, and to it disturbed digestion, acidity of the stomach, and defective condition of the excretions, with general *malaise*, were supposed to belong. On the other hand, however, it should be borne in mind that the commencing period of rickets was entirely unknown before the discovery of rachitis of the skull by *Elsässer* in 1843, and that most of the signs of the so-called prodromatory stadium are now seen to be prolonged far into the disease itself.

Rickets is a visible and comprehensible disease, and it is therefore necessary to investigate more minutely the alterations of the individual parts of the body which result from it during life.

A.—*RACHITIS OF THE SKULL.*

Rachitis of the skull, with its peculiar phenomenon—softening of the occiput—was discovered by *Elsässer*. It is worthy of remark that, previous to the publication of *Elsässer's* work, no physician had any idea of this extensive morbid condition of the occiput, though it is one easy of examination and of detection. *Neumann*, for example, says that the bones of the head never soften through rachitis; on the contrary, they often grow at the expense of the other parts of the body. *Miescher* says that all the bones soften except those of the head. Various other remote alterations are indeed likely to originate about it, such as increased growth above the usual dimensions. *Schnitzer* and *Wolff* say that the bones of the skull never soften; they even grow, apparently at the expense of all the other parts.

The following are some of the alterations which occur about the rachitic skull:

The anterior fontanel, which, in normal children, closes at the latest at the end of the second year, remains open three to four years, and may even remain cartilaginous up to the sixth year. The serrated suture, which otherwise we find closed by the end of the first year, is frequently still ununited in the third year. The coronal suture, instead of being united in four months, remains open at the end of two years, and the lambdoidal, instead of being closed at three, is still open at the end of fifteen months. *Rufz* has instituted accurate measurements of the skull, and found that the longitudinal and the

transverse measurements exhibit but slight deviations from the normal; but the peculiar angular projection of the protuberances of the frontal and parietal bones robs the sinciput of its usual globular form, and gives it a quadrangular, clumsy shape (*tête carrée*).

After recovery from the disease, a depression usually forms along the course of the coronal suture, which gives to the sinciput, when seen from above, the form of a calabash, and is due to an hypertrophy of the frontal bones. Numerous depressions and elevations in general take place during rachitis, which opens a wide field of research for cranioscopy.

The soft occiput is met with in children from the third month on, but is seldom seen in those who have passed the second year. No constant prodromata are observed. Many children may have, indeed, been previously subject to a bronchial or intestinal catarrh; others, however, have enjoyed the best of health, and, up to the appearance of the craniotabes, were well nourished, fresh, and hale.

The disease begins with profuse cephalic perspiration, which often soaks through the pillow, and a nocturnal restlessness, increasing gradatim, becomes noticeable at the same time. Children, who otherwise slept uninterruptedly for several hours, wake up every quarter of an hour crying, and rub and bore the head into the pillow. Changing the posture of the head quickly tranquillizes them, but only for a short time. The incessant rubbing of the head on the pillow produces a complete alopecia of the entire occiput.

After some time the whining and discontentedness grow worse, extending even to the daytime, and attentive nursery-maids soon notice that the discomfort is subject to the varying positions of the head. The patients cry, and constantly bore the head when they are kept horizontally, or on the arms while being fed, or put to sleep, but soon become quiet when they are raised up and the occiput is relieved from all pressure. They then take their food with the greatest comfort, and prefer also to lie with the face resting upon the nurse's shoulder, and the occiput entirely free. Older children occasionally quickly turn over in bed on the belly, and lie with the forehead pressed into the pillow.

The feeble growth of the hair in general, and the alopecia of the occiput, are very noticeable, and, on closely inspecting the skull, the occiput is generally found flattened and the protuberances more angular than usual. For the purpose of a more accurate manual examination, the occiput should be taken upon both hands and felt of with the ends of the fingers.

I examine the whole posterior region of the head, from the lambdoidal angle to the mastoid process, *twice* carefully by the ends of the

fingers. The first time, for the sake of precaution, I exercise only a very mild pressure with the flat, open fingers, so that, in case large, very soft places exist, no great violence may be done to the unprotected brain. The second time, I bend the fingers at the point a little, and press forcibly upon every part of the occiput and parietal bones. By this procedure even the mintuest point of attenuation marked by depressibility may be discovered with certainty.

The soft places generally vary in size from that of a lentil up to that of a bean, and are found in the vicinity of the lambdoidal and posterior portion of the sagittal sutures, and sometimes encroach upon the sutures. The external occipital protuberance only is always spared. The diseased parts of the bones are elastic, their original convexities may be converted into equally as great concavities, and, when pressed, yield like a card when laid across a hollow, or like an inflated dried bladder. The pain attending a careful examination is not very great.

The most frequent complications of this affection are spasms of the varying groups of muscles. The most dangerous of these is spasm of the glottis, whose undoubted yet by no means physiologically explained connection with craniotabes has already been discussed in detail on page 274.

Besides attacking the occiput and the sinciput, rachitis invades the jaws. The teeth cease to grow, so that the patients get to be twelve and eighteen months old before they cut the first incisor teeth. After these have finally appeared, they turn black, and, for want of enamel, crumble down. When the enamel is totally wanting, the whole tooth down to the margin of the gum will disappear; sometimes it is only deficient on the apex, and the blackness is then restricted to that point. As the disease disappears before the second dentition commences, these phenomena are not observed in the permanent teeth.

Deficiency of the enamel, now and then met with in older children, is, according to the statements of some authors, due to the use of mercurial preparations, especially calomel. Should it actually be statistically demonstrated that the majority of these children had taken calomel, it would, very properly, in future, much restrict the use of this medicine.

B.—*RACHITIS OF THE THORAX.*

Glisson and his contemporaries correctly recognized the rachitic process, in the condition known as pigeon-breast, and subsequent authors devoted much of their time and attention in investigating the manner of its origin; we therefore have much more explicit data concerning rachitis of the thorax than of craniotabes.

It usually comes on somewhat later than softening of the occiput,

and many children who fortunately escaped the latter, and are already being carried about upright, are attacked by rachitis of the thorax. Perceptible alterations are seldom observed in children under six months, while craniotabes may be present as early as the third month of life. The statement made in some of the text-books, that the pigeon-breast occurs from the first to the fourth year of life, is to be understood as meaning that children so old as four years may be observed with this disease. But, after the completion of the first dentition, rachitis never comes on in a child hitherto perfectly healthy.

The first symptom of rachitis of the ribs is a marked pain on touching or pressure of the thoracic walls. Nurses say that "the child cries every time we raise it up, if ever so tenderly." Usually such statements are not much heeded by the physician, because most of them are based upon prejudice and incorrect views. The frequency of these complaints, however, struck me long ago, and I have convinced myself that it is by no means a rare occurrence that children between five and ten months old suddenly cry out in pain when they are grasped with both hands under the axillæ and lifted up, and, as soon as they are laid down, become tranquil again. Nay, more, it is not even necessary to lift them up; slight pressure with the finger in the axillæ or on the thoracic walls generally, suffices to produce pain. If such a child is tenderly raised with one hand under the pelvis, and the other supporting the neck, it will remain as quiet as if it had been lying on the pillow, and in this manner, also, its bedding may be changed without giving it any pain.

At this time, little or no hypertrophy can be felt at the sternal end of the ribs, the boundary between the costal cartilages and the bone. The sternal ends of the ribs do not begin to appear bulbous and hypertrophied, so as to be detected by the finger, and later also by the eye, till after several weeks. Thus two uniform rows of buttons, the so-called rachitic wreath, appear on both sides of the thorax at a point corresponding to the end of the costal cartilages. These buttons, so palpable from without, project still more internally, forming large angular tubercles which encroach upon the cavity of the thorax.

The thorax always becomes deformed in those cases where these hypertrophies have existed for some time. The sternum, which likewise undergoes softening, is pushed off more and more from the spinal column, and arches outwardly; the xiphoid cartilage becomes extremely movable, and, projecting, forms a deep pit in the scrobiculo cordis. In the severest grades of pigeon-breast, the costal cartilages immediately behind the sternum run straight backward to meet the elongated transverse processes of the spinal column, and thus, at their anterior ends, the ribs form a concavity instead of a convexity.

The diameters of the thorax become smaller from side to side, and larger antero-posteriorly, as is shown by the delineation, Pl. V., Fig. 2. The transverse diameter of the thorax assumes the shape of a pear, whose apex is supposed to be at the sternum. The rachitic wreath is found chiefly from the second to the eighth rib, the false ribs are forcibly pressed outward by the liver on the right, and by the stomach and spleen on the left side. The abdomen, in consequence of the constant tympanitis, and a shortening and curving of the spinal column, is tumefied, and of globular form, and much encroached upon by the distorted thorax. The spinal column is curved most during the sitting posture, and the globular shape of the abdomen is also on that account most striking in this position. When these children are laid upon the belly, and in this position raised up, the external curvature of the spinal column disappears entirely, and assumes again its normal form. In neglected cases, and where the rachitis has existed for several years, a permanent arching—not an angular curving—of the dorsal vertebræ, laterally and posteriorly, may take place.

The origin of the pigeon-breast is explained, in part, by pressure of the atmosphere upon the soft ribs, and, in part, by the traction of the diaphragm, for which they serve as points of attachment. Having lost their firmness, the ribs are no longer able to withstand the constant dragging inwardly by the diaphragm.

From rachitis of the thorax there originate (1), *an alteration in the curve of the ribs;* and (2), *an arrest in the longitudinal growth of the ribs*, a still more serious result, which inevitably diminishes *the pectoral space*, and promotes that disease of the lungs known as acquired atelectasis, as has been already conclusively shown on page 299.

The prognosis depends exclusively upon the affection of the lungs. When a great portion of them is involved in the atelectic process, and has become impermeable, then, of course, a serious catarrh in the remaining normal tissue suffices to induce labored breathing, and even dyspnœa, suffocating attacks, and death. In this complication, in fact, we have the usual cause of death in rachitic children, as *Romberg*, *Guersant*, and others, have remarked.

C.—*RACHITIS OF THE PELVIS AND OF THE EXTREMITIES.*

The pelvis does not become deformed before the rachitic child is able to walk, and then it is the result of scoliosis, or of an inequality of the lower extremities, after the manner of a distortion of the pelvis in coxarthrocace. The important consequences of this alteration in the female are discussed sufficiently in detail in the standard works on obstetrics.