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A CASE OF CONGENITAL DEFORMITY OF THE UPPER AND LOWER EXTREMITIES AND OF THE SPINAL COLUMN.

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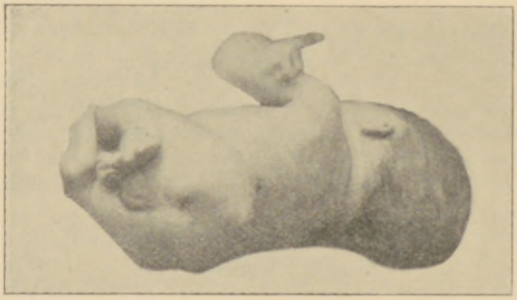
ON September 22, 1891, I was called to attend Mrs. C., who was reported to be suffering from an attack of "cramps." I found my patient to be a young woman, eighteen years of age, of small stature, thin in flesh, pale and anemic, and of a highly nervous temperament. I was told by members of her family that she had been marrried only four months and had not had her menses during that time.

After observing the patient a few moments, it was evident that she was in the first stage of labor. Upon digital examination I found the os partially dilated and the occiput presenting. The abdomen was not as large as is usual in pregnant women at full term. Labor progressed normally for about one hour and a half, when the uterine contractions began to grow weak and finally almost ceased. One hour later, when I returned to my patient, I found the labor-pains quite severe and following one upon the other in close succession; from this time on labor advanced normally to delivery, four hours and twenty minutes after I was first summoned to the case.

A boy-baby with numerous deformities was born. These consisted in absence of the lower end of the spinal column, the coccyx, and the protruding backward of the

lower end of the sacrum. Double club-foot was present, double club-hand, displacement of both ankles, imperfect knee-joints, and left hip-joint. There was also imperfect development of the wrist-joints and elbow-joints, flat-feet, flat-hands, imperfect external ears, a double great toe on each foot to the distal joint, with double nail. The fingers were normal and their joints perfect; the great toe on each foot was unusually long, fully twice its normal length; the other toes were perfect. The child was a strong, fleshy, vigorous, and healthy infant.

FIG. 1.



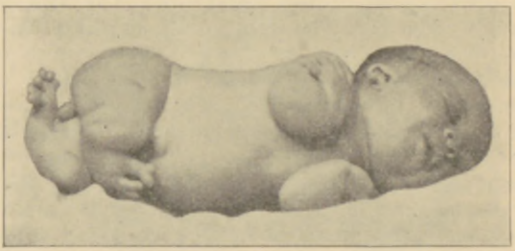
Perhaps the most striking deformity was the absence of the coccyx and the pointing backward of the sacrum. This deformity is well shown in Fig. 1. It was not a spina bifida; the prominence seen above the anus was of bone, and resistant to the touch, and not of that smooth elastic nature found in tumors caused by incomplete closure of the bony canal in the spinal column. It contained no fluid, and could not be reduced in size by pressure. The right foot was in the most extreme form of talipes equino-varus; the sole of the foot pointed upward and rested on the abdomen. The foot was flat, the plantar fascia and internal lateral ligaments were

shortened, and the tibialis anticus and posticus muscles were contracted. There was marked displacement of the tarsal bones; the astragalus was tilted downward, the scaphoid was displaced inward and downward by the action of the tibialis posticus, and the tubercle on this bone was very prominent; there was in addition marked rotation at the astragalo-scaphoid and calcaneo-cuboid junctions. The displacement of these bones gave the foot a short, flat appearance, somewhat resembling the human hand. The os calcis was rudimentary, and in consequence the heel was less prominent than normal. The articulation at the ankle-joint was imperfect, owing to the displacement of the astragalus. The foot was strongly adducted. The right knee-joint was displaced inward at a right angle, the tibia and fibula were shorter than normal and curved inward, and the patella was rudimentary on the right side. The femur was normal and in the socket at the acetabulum. The right hip-joint was perfect in development, and admitted of free motion in all directions. The left foot was in extreme talipes equinus. The gastrocnemius and soleus muscles were shortened, the tendo Achillis was tense, so that flexion of the foot was impossible; the foot was slightly adducted. The sole of the foot rested on the scrotum and buttocks, from the strong flexion of the limb at the knee and thigh. The left knee was strongly flexed; efforts at extension were met by bony and ligamentous resistance. The left hip-joint was also in extreme flexion, and could be extended only to an angle of 45°. The left femur was normal in length and development. The flexion at the hip-joint was due to a contracted condition of the flexor muscles of the thigh.

The deformities were alike on both sides in the upper extremities, and resembled very much those found in the lower limbs. The humerus was normal in length and perfect in development on both sides. The shoulders were perfect and admitted of free motion. The deformities of

the upper extremities are well shown in Fig. 2. The forearm was tightly flexed upon the arm and could not be extended beyond an angle of 35°, from contraction of the biceps muscle. The radius and ulna were shorter than normal, and curved inward; the ends of these bones were rudimentary. Efforts to extend the forearm at the elbow caused a creaking noise, especially when forced beyond an angle of 35°. The wrist-joints were rudimentary, the hands were adducted. The palms of the hands rested on the arms in the deltoid region; there was

FIG. 2.



slight motion at the wrist-joints. The fingers were normal in length, and the phalangeal joints perfect. The external ear was marked by the absence of part of the lobule. The head and trunk were perfect in form and development, except for the absence of the coccyx.

The infant cried lustily after birth, was well nourished, and gave every promise of living if properly cared for and nourished.

The father, when he saw the hideousness of the deformity, and being desirous also to conceal the birth of a child so soon after his marriage, placed it, contrary to my wishes and advice, in a foundling-home in Philadelphia. The physician of that institution reported its death as due to debility when it was twenty days old.

Medical literature is replete with instances in which vivid mental impressions are supposed to have produced a direct effect upon the development of the fetus. Mrs. C. gives a history of having received at least two severe mental shocks during her period of gestation—one seventy-one days after conception, and another three months before parturition. On February 25, 1891, while employed in a paper-box factory, she saw the clothing of a friend caught by a revolving shaft, which drew her to the floor, and as she fell her limbs and arms were twisted about the shaft. Some of her clothing was torn off, and there was considerable blood lost by the tearing out of part of her hair. Mrs. C. was close by when the accident occurred, and was very much frightened, and did not fully recover from the shock for several months.

She was, as I have already said, naturally of a weak, nervous temperament, which condition was, no doubt, augmented by her having conceived out of wedlock and having to support herself by working in a close, dusty room in a box-factory. Her surroundings were not calculated to produce that cheerfulness and equanimity of mind desirable for pregnant women; on the contrary, her weak body and agitated mind were well calculated to produce the result in her offspring that has been here shown, if sufficient emotional excitement were brought to bear upon her. She gives February 25th as the day of the fright. It was the birthday of a lady friend, who visited her in the factory that day and saw the accident with her. Supposing the child to be a full-term baby, if we count back 280 days, it would bring the day of conception on December 16, 1890. This was seventy-one days before February 25th, the time of the fright.

Mrs. C. gives the history of having had a second fright in June, 1891, about six months after the beginning of pregnancy. At this time her own clothing was caught by a revolving belt, and part of it torn from her body. This accident does not seem to have made as

vivid an impression upon her mind as the first, as she does not recall it in all its minutiæ, as she does the one of February 25th.

I wish to mention in this connection that the woman for three or four months prior to parturition had been in the habit of lacing herself very tightly, so as to conceal from friends and relatives the near approach of her maternity. She had so far misled her relatives that they administered anodynes for her "cramps" prior to sending for me. Authorities on this subject do not believe that tight lacing has anything to do with the production of deformities.

On November 19, 1892, fourteen months after her first confinement, I delivered the woman of a perfectly formed male child. This second child has since developed a strumous diathesis, and at present has enlarged suppurating glands in the neck.

The husband has lately died of tuberculosis of the lungs, with which disease he had suffered for five years or more. His family history is tuberculous. The mother's relatives are all healthy, and none is deformed.

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