THE PATHOLOGY
OF CONGENITAL RING CONstrictions*

by

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There is no agreement about the cause of the constrictions at birth on various parts of the human body. This paper suggests that they are due to the digits, limbs or the body itself penetrating to various degrees the amniotic membrane, with consequent concentric pressure on the developing tissues. The conditions necessary for a final testing of this hypothesis are laid down.

Characteristics of Ring Constrictions

1. They increase in frequency with their nearness to the extremities of the body, being very common in the fingers and toes, less common on the limbs and extremely rare on the body. I have seen one case with a typical one right round the lower thorax, but have not heard of another.

2. It is common to find two or more digits fused together at their tips, with a ring constriction round the fused region, while their proximal parts are separate.

3. The constrictions may vary from a slight groove in apparently normal skin and subcutaneous tissue to such a depth that the part distal to them may fall off, either before or after birth. In the deeper ones ulceration of the bottom of the groove at birth is usual. Intermediate depths may cause atrophy and paralysis of the distal parts, with or without lymphatic oedema. This oedema subsides after excision of the ring and joining of the skin on either side of it.

4. Occasionally there are constrictions at two levels on a limb, for instance, round the tips of the toes and also round the ankle.

5. There may be an acute bend of the bones at the site of the constriction. I have seen this only in the leg.

6. Club-foot and other deformities of the feet are frequently associated with constrictions.

7. In certain cases strips of tissue resembling necrosed amniotic membrane are found in the constrictions. This only occurs when they are very deep, with ulcerated bases.

8. It has been noted in many cases that the amnion is irregular and torn or shredded, apart from showing 'amniotic bands' in some cases, but not in all.

9. Apart from the occasional talipes moulding of the feet which has been mentioned, the children are normal, with no sign of excessive mechanical or hydraulic intra-uterine pressure.

10. An important case in my argument, though there were no constrictions on the child, is a surviving girl from an extra-membranous pregnancy. She is now a normally active child though she had a club-foot and suffers from Turner's syndrome.

I suggest that this whole picture can be explained by the assumption that these children have got their digits, their limbs, half their body, or in the case of extra-membranous pregnancies, their whole body, through or into holes in the amnion. To illustrate the mechanism one can cut a round hole in a piece of rubber sheeting and thrust a wax candle through this hole so that the rubber is firmly contracted on the wax. If the wax is then softened by heating in hot water the contracting rubber will mark it in a ring constriction very like those found on the body. Obvious modifications of the experiment will reproduce the typical fusions of digits at the site of the constriction.

The shreds of necrotic amnion found in some ulcerated constrictions can be reasonably explained as the products of a mutual compression between the constricting ring of the perforation and the limb gripped by it; pressure high enough to cause the sloughing of the foetal skin must also be high enough to kill the amnion in immediate contact with it. As the gripped limb is torn out it brings the necrotic ring with it.

Constriction rings at different levels on a limb could be the results of a limb which had originally penetrated only a short way into the amnion subsequently being thrust deeper.

The importance of the case of extra-membranous

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FIG. 1.—Newborn baby with congenital constriction of the left leg. Note oedema below the constriction.

FIG. 2.—Hypothetical diagram of the intra-uterine conditions that may have been responsible for the constriction shown in Fig. 1. The amnion is represented as a transparent sheet by fine parallel lines.

FIG. 3.—Case shown in Fig. 1 after (a) excision of the constriction; (b) osteotomy on right-angle bend of the tibia and fibula at the site of constriction; (c) correction of club-foot moulding of both feet. The child is now, at the age of 7, very strong and active on her feet.

Pregnancy (Lawlor, 1949) is that if a child can escape wholly from the amniotic case it is reasonable to assume that partial escapes may also occur, and that they would be much more frequent; the frequency diminishing as the degree of the escape increases.

Streeter (1930) has suggested that the rings are due to intra-uterine ulceration in a ring form, resembling a tropical disease called ainhum. Objections to this are: (1) Many of the slighter constrictions show no evidence of ulceration ever having been present. (2) This ulceration could not cause the fusing of the tips of the digits since the raw areas, which alone could fuse, are at the bottom of the grooves and cannot meet each other. (3) It cannot explain a ring round the body; one cannot imagine an annular degeneration of this kind there. (4) It gives no explanation of the shreds of necrotic tissue in the deep clefts; processes of ulceration never produce anything like these.

Many authors have suggested amniotic bands as the cause (Lennon, 1947; Grob, 1957). A few experiments with a limb of modelling clay and a 'band' of any kind, say a piece of tape, will show that it is quite impossible to produce any ring marking in this way.

It is important to note that in my opinion the condition, seen most frequently in the forearm, in which the limb ends suddenly, usually with rudimentary digits, is a totally different one, due to simple failure of development.

Various points remain to be investigated along the lines I have suggested. The first essential is the
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Fig. 4.—Similar case to that in Fig. 1.

Fig. 6.—Typical constriction rings, fusions and amputations of toes.

Fig. 5.—Typical congenital constrictions of the fingers, showing fusions and amputations.

Fig. 7.—Constriction ring of the left leg so severe that the distal part became gangrenous and dropped off soon after birth. Note club-foot deformity of the other side.

preservation and careful study of the whole of the membrane in a case of the sort. Unfortunately the placenta is seldom studied, and the membranes almost never. It would be necessary for proper examination to have some sort of a mould about the size of the folded foetus, upon which they could be pinned and fixed for investigation. Then there is the stage of pregnancy at which the penetration of the amnion occurs to be determined. The common fusing of the digits suggests that this is very early, before they have separated.

REFERENCES