CONGENITAL CONSTRICTION BANDS IN SINGAPORE

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This curious anomaly affecting especially the digits and the rest of the limb where it is more marked below the elbow or the knee has not been reported from this part of the world. This report consists of 30 cases that have been collected in this hospital from the orthopaedic records of the last twelve years. However only over the last few years has the anomaly been recognised sufficiently to record it as such. In many of the earlier cases an initial diagnosis of syndactyly or merely congenital abnormality was made. There are obviously therefore many missed cases. Moreover a large number of affected individuals would not have sought medical attention. As it is routine practice now in the University unit to index all cases and keep photographic records, the cases seen over the last three years reflect fairly accurately the incidence of this condition.

RACE AND SEX INCIDENCE

RACE	MALE	FEMALE	TOTAL
MALAY	11	11	22
CHINESE	4	3	7
INDIAN	1		1
			30

Fig. 1. To show the race and sex incidence in the 30 cases.

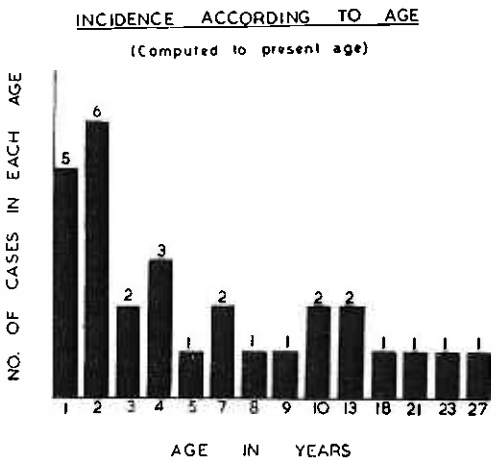

Fig. 2. To show the incidence at each age computed to the present age.

Figure 1 shows the race and sex distribution of the 30 cases. The present population of Singapore is 1.8 million with the Malays forming 14% of this population. Figure 2 shows the incidence according to age and it can be seen that most of the cases were seen over the last few years. Some older children were also seen during this period. For the years 1961 – 1963 the incidence at birth in the total population is 1 in 15,000 live births but for the Malay population this is 1 in 4,000 live births (and most likely it is higher). This I believe is a world record.

Until recently all cases of congenital absence of a limb have been bunched together but really there are two distinct varieties.

In aplasia of the limb there is present a stump to which rudimentary digits are attached (Fig. 3). There is no constricting band associated with this condition. In true intra-uterine amputations from congenital bands it is the digits that are commonly involved (Fig. 4). In addition to terminal absence of a part there are present constricting bands in other parts of the limb either in the same digit at a proximal level (Fig. 5) or in the other digits (Fig. 6) or in the limb at a higher level (Fig. 7) though usually below the knee (Fig. 8) or the elbow joint (Fig. 9). There is also fusion of the digits — a variety of syndactyly (Fig. 6). Again supernumerary digits showing constrictions at the base are not to be confused with congenital constriction bands. (Fig. 10).

The constricting grooves are usually circumferential, hence the term annular, but only a part of the circumference may at times be involved. The depth varies from a broad shallow depression to one where the whole limb is so severely constricted that the distal portion becomes gangrenous and sloughs away (Fig. 9). In the digits a little interference with the blood supply causes gangrene and this is why I feel they are generally amputated (Fig. 5). In a number of cases the terminal dead portions have been found to be attached to the remainder of the digit in the form of filaments. Limbs have been delivered separate from the infant after it has been born.

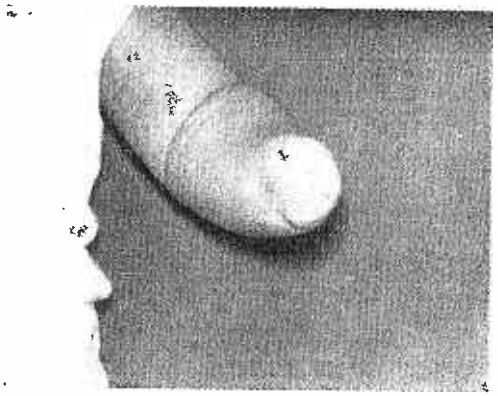

Fig. 3. Showing aplasia of the hand. Note the rudimentary digits at the end of the stump.

SITE INCIDENCE

UPPER LIMBS : 34	FINGERS : 30
LOWER LIMBS: 35	TOES : 34

Fig. 4. Shows the site incidence. Note that the fingers and toes were commonly involved.

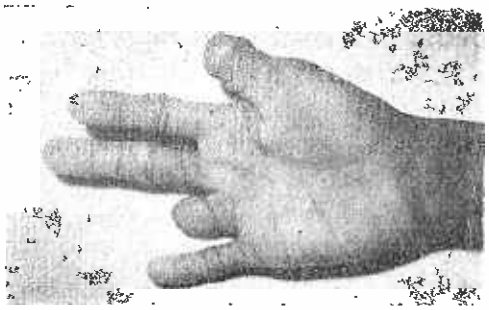

Fig. 5. Note the amputated thumb and ring finger with grooves on them.

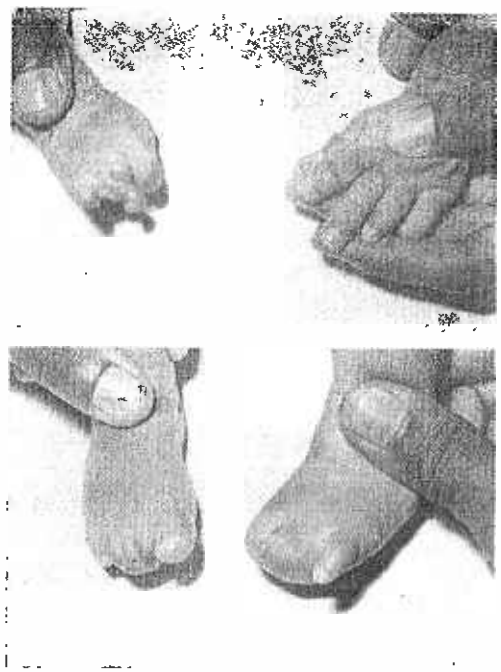

Fig. 6. Showing in the digits: amputation, syndactyly and annular grooves.

Fig. 7. Patient whose hand is shown as a close up in Fig. 5. Note involvement of the right foot and the groove in the leg.

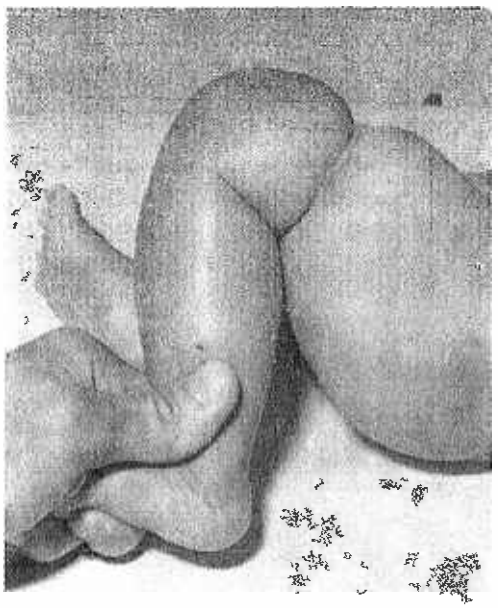

Fig. 8. A band around the thigh — a rare type.

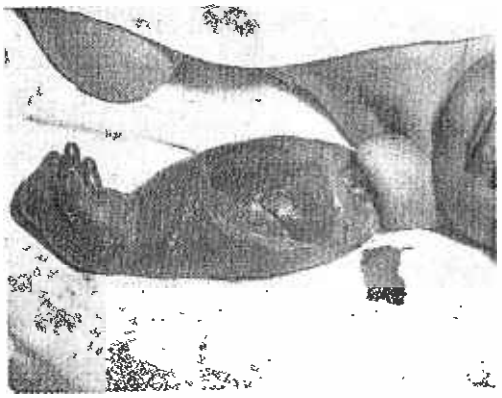

Fig. 9. Constriction above the elbow (rare) leading to gangrene at birth.

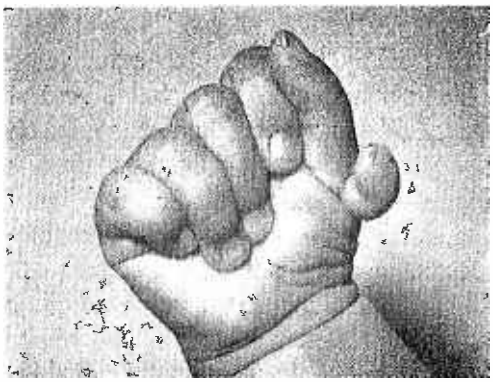

Fig. 10. A supernumerary thumb with constriction of the base — not a case of constriction band.

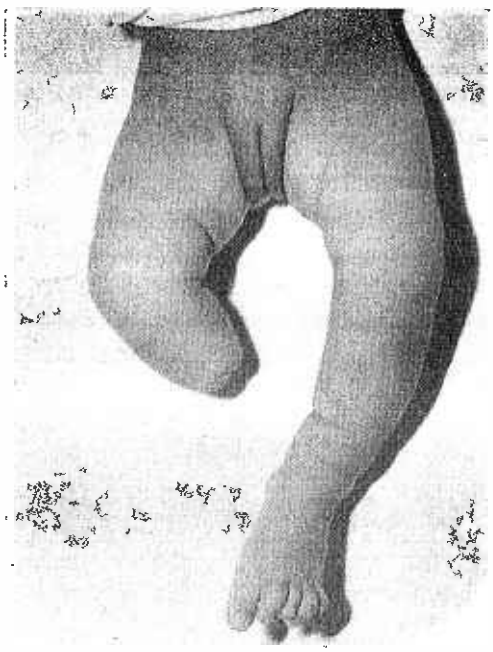

Fig. 11. Congenital amputation from constriction band. Note the involvement of the toes in the left foot.

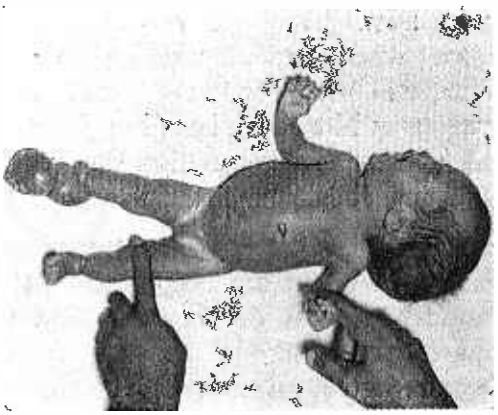

Fig. 12. Marked bands of the lower limbs. Note that only the dorsum of the right foot is involved and that the left middle finger is amputated.

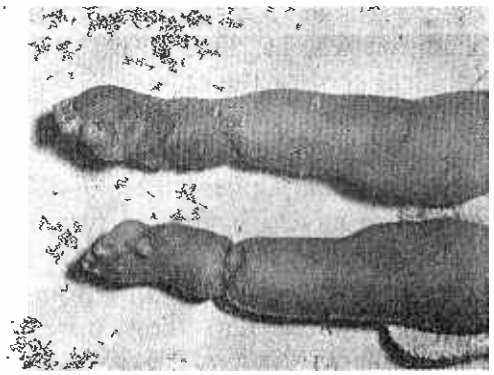

Fig. 13. The lower limbs of the case shown in Fig. 12 after two Z plasties to the front of the right leg and foot. The back of this leg and the left leg have not been done yet. The left big toe is also involved.

Fig. 14. The condition affecting the toes and the talipes equino-varus of the foot are shown.

In some cases the whole amputated limb has been missing and was probably lysed and absorbed when the amputation took place early in fetal life. (Fig. 11). Limbs that are gangrenous at birth have been seen locally (Fig. 9) and in these the amputation was completed by a formal operation.

In the majority of cases the cicatrising bands extend up to the deep fascia only and the muscles, tendons, deeper vessels and nerves are spared. Anaesthesia distal to the groove has been reported. Excision of the scar underlying the groove and suture, will improve the oedematous state of the distal portion but a recurrence is usual, though this is not severe. It is best to do multiple Z-plasties as these will give excellent results. It will be wise to do this in stages tackling half the circumference at one sitting (Figs. 12 & 13).

Before Streeter's classical paper in 1930 this condition was ascribed to constriction by the umbilical cord or the amnion. The former concept is ludicrous but pieces of amniotic membrane have been found in the grooves. These are secondary phenomena and in most cases they are merely hyalinised fibrous tissue or macerated sheets of epidermis. Streeter in a dissection of fetuses showing intra-uterine amputations showed that these were due to focal areas of dysplasia. These were circumscribed areas of defective tissues which were viable only for the first few weeks of development. These defects were seen in the mesoderm especially in the subcutaneous tissue. Bagg in 1929 reporting the effects of radiation in mice prior to conception showed that a large number of the progeny had congenital limb abnormalities such as club feet, syndactyly, polydactyly and congenital amputations.

These features were due to germplasm deficiencies and were inherited as recessive traits in subsequent generations. A study of the fetuses in utero showed that early in the development of the limb bud, areas of perivascular lymph stasis occurred in some parts especially the dorsum of the foot-plate. Blood entered these spaces and a part of the limb was thus destroyed. It was this that lead to the deformities mentioned. In this connection it is interesting to note that though I have seen bands on the dorsum of the foot I have not seen it across the sole of the foot (Figs. 12 & 13). Moreover the incidence of club feet is very high in children showing constriction bands (Fig. 14). Eleven of the Malay and two of the Chinese cases had club feet of varying degree of severity.

Though genetic factors may play a part it is possible that dietic insufficiency too is important. Riboflavin deficiency can cause skeletal malformations in animals and Duraiswami showed that riboflavin and nicotinamide injections could protect the chick embryos against the harmful effects of insulin injections. In the 10 Malay, 4 Chinese and 1 Indian cases seen by me, generally the children came from poor homes. It is usually the mother who will be malnourished in these homes. The factors that cause these anomalies must operate during the first trimester when the limb bud shows rapid growth. There was no history of X-irradiation before or in the first half of pregnancy. In one case there was a history of maternal ill-health and in one a definite attempt to abort the fetus by drugs and massage. There was no history of a similar affliction in any other member of the family or in relatives except in one Malay case where the maternal grandmother was said to be similarly afflicted.

BIRTH ORDER IN 10 MALAY, 4 CHINESE & 1 INDIAN CASES.

BIRTH ORDER Position in family	1st	1st	3rd	4th	4th	5th	5th	5th	5th	6th	6th	6th	7th	8th	9th
NO. of CHILDREN in family	1	2	3	4	8	5	5	6	7	6	6	6	7	9	9

Fig. 15. Shows the birth order in the cases personally reviewed. Most of them are among the later offspring.

A few of the mothers ascribed the defect as a visitation for cutting off crab's legs by a parent during the pregnancy. The birth order is striking (Fig. 15). They appear to be among the later births. In acrocephalosyndactyly, Blank showed that the propositi were among the later births. This might lend some support to a germplasm defect.

SUMMARY

30 new cases of congenital constriction bands are presented. The etiology is obscure though the pathology is better understood. The high incidence in Malays is striking.

ADDENDUM

Two further cases have been seen since the preparation of this paper — one is a 5

year old Indian girl and another is a female Malay infant.

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