

Congenital malposition of the feet and hands may occur in consequence of faulty position, or undue intra-uterine pressure, without defective development of the skeleton,

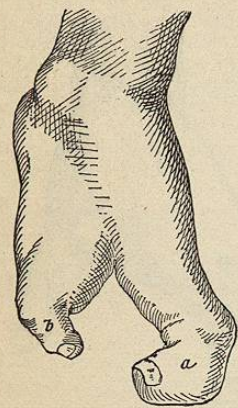


FIG. 4695.—Deformed Foot (Peropus). a, b, Great and little toes. (Otto.)

all types of club-foot or club-hand being observed. In other cases the distortion of the distal segment is associated with defective development of the bones of the forearm or the leg, as of the ulna or radius with talipomanus, or of the tibia with talipes. In the consideration of spina bifida (page 697) attention was called to the frequent occurrence of club-foot with such malformations of the spinal cord. In such cases the talipes results from the loss of balance in the muscular forces acting on the limb in consequence of impairment of the nerve supply to the muscles from the spinal cord. The causes leading to the various foregoing malformations of all degrees are chiefly those which induce abnormal pressure upon the developing limbs, whether the unfavorable influence be exercised directly by the amnion, or by inadequacy of the space within the uterus. Amniotic attachments or constricting investments are undoubtedly prolific sources of these malformations. Barwell,¹³⁶ however, insists that defective limbs are less often the result of constrictions than usually assumed. The histories of many cases, moreover, point strongly to heredity as an additional factor in their production. Ebstein¹³⁷ has reported a remarkable example of syndactylism of the fingers and toes through five generations. In other instances it is difficult to account for symmetrical malformations profoundly affecting all the limbs without recognizing an inherent lack of primary development, apart from the dwarfing influences of mechanical forces.

Congenital Redundancies of the Extremities.—These malformations include the frequently observed supernumerary fingers and toes and the rare duplication of the hands or feet. The few instances of reputed doubling of the entire lower limb that have been recorded must be viewed with doubt, since it is highly probable that these cases are to be regarded as double monsters.

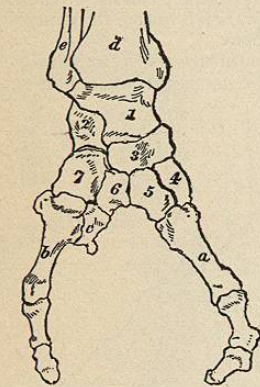


FIG. 4696.—Skeleton of Preceding Foot. (Otto.)

Polydactylism exhibits all degrees of doubling of the digits, from the scarcely evident partial cleavage of the distal phalanx to the production of a complete two-fold quota of fingers and toes. The presence of ten digits, however, is very exceptional; that of seven or eight is also rare, the most common addition being of only a single supernumerary finger or toe. The position of the latter is usually marginal and about one and one-half times more frequent on the lateral (radial or tibial) than on the median (ulnar or fibular) side. The thumb is oftener doubled than the great toe. The doubling may involve only the phalanges, or the cleavage may begin with the metacarpals or metatarsals. When an entire, comparatively well-formed additional digit is present, it may articulate with the metacarpal or metatarsal of its neighbor, to join directly with the carpal or metacarpal series. More rarely the latter may be

also augmented in number to provide special bones and attachments for the extra digit. All possible combinations occur in the relations of the supernumerary parts with the usual digits, especially in conjunction with an associated syndactylism often coincidentally present. When very imperfectly developed, the additional finger or toe may be so rudimentary as to be merely a knob-like appendage possessing only integumentary attachment. The extra digits may occur only on a single limb, on both hands or feet, or on all four extremities, although their position is by no means always symmetrical.

The etiology of polydactylism has elicited much discussion, and many theories have been advanced in explanation of these frequent malformations. The assumed causes may be grouped under two headings—external and internal. Foremost among the former are the mechanical impressions due to amniotic cords or folds; among the latter are heredity and excessive differentiation. The presence of opposing amniotic folds is regarded by some authors as the most important factor in producing supernumerary digits which are regarded as arising in consequence of cleavage of the primary anlage, from each portion of which is formed a more or less perfect finger or toe. That in a certain number of cases amniotic folds are probably accountable for the fission resulting in supernumerary digits must be admitted, but the acceptance of such mechanical disturbance as the chief cause of such malformations fails to account for the acknowledged influence of heredity, which is particularly conspicuous in polydactylism. The remarkable case of Muir,¹³⁸ in which supernumerary digits were present in five consecutive generations, leaves little doubt as to the importance of these unknown influences in the production of polydactylism.

That the presence of amniotic folds is not a necessary condition has been shown by Tornier,¹³⁹ who pointed out that polydactylism is particularly frequent in amphibian embryos in which the development of an amnion never takes place. Neither is the assumption convincing that the influence of heredity is not directly exerted upon the limb-anlage, but upon the amnion, in consequence of which a peculiar relation of the amnion to the developing limb is established, resulting in digital fission. It seems scarcely plausible that hereditary influence suffices to arrange amniotic bands with the symmetry necessary to produce supernumerary digits upon all four limbs, as sometimes is the case. Since the evolution of the mammalian limb is still

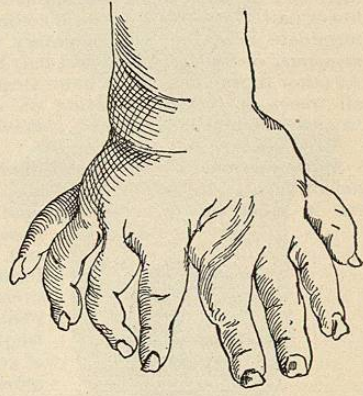


FIG. 4697.—Polydactylism of Hand. (Ziegler.)

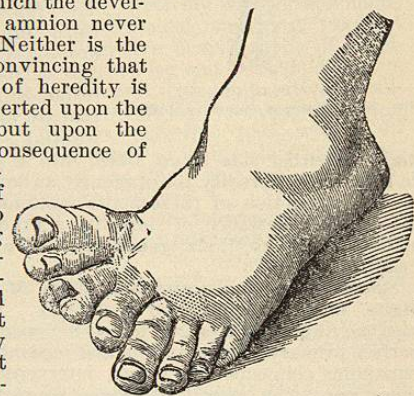


FIG. 4698.—Polydactylism of Foot. (Johnson.)

shrouded in much uncertainty, we may admit our ignorance concerning the exact rôle played by heredity in producing polydactylism; it is not unreasonable, however, to assume that the impression in some way influences the growth processes which normally result in the differentiation of five digits.

MALFORMATIONS OF THE INTEGUMENT.—The skin is the seat of a variety of congenital abnormalities, some local and others extensive, which depend upon unusual conditions of (1) the epidermis, (2) the cutis and subcutaneous tissue, and (3) the appendages—glands and hair. Since not a few of these congenital abnormalities are caused by pathological lesions more than by errors of development, their consideration falls within the province of dermatology rather than of teratology; such will, therefore, here call for but brief notice.

Abnormalities of the epidermis include more or less general thickening of the epithelial tissue of the integument, which may constitute a horny stratum, several millimetres in thickness, broken only by the furrows caused by the folding and movements of the members of the body. This condition, known as *hyperkeratosis congenita* or diffuse keratosis, is usually characterized by loss of hair and obliteration of sebaceous glands. Scale-like patches of hypertrophied epidermis distinguish the abnormality termed *ichthyosis congenita*, a condition in which, when extensive, almost the entire body of the fetus may be covered by irregular areas of greatly thickened epidermis, so that the integument presents a scarred and fissured appearance. When the horny investment of the extremities is very marked, the shell so formed may act as a mechanical restraint to the normal development of the hands and feet, which may, therefore, be defective.

Anomalous pigmentation includes excessive and defective development. The former condition is represented by the smaller or larger colored spots (*naevi pigmentosi*), often very numerous, which may be smooth and unelevated, or raised above the level of the skin and frequently beset with hairs. The color of the pigmented areas varies from a pale to dark brown or black, the epidermis being usually of normal thickness. Deficient pigmentation or *albinism* of the skin and hair, usually also evident in the iris, choroid, and retina, may be partial or complete. In the latter case the abnormally light-colored skin, white hair, pink irides, and red pupils (due to the vascular reflex through the pigment-free retina and choroid) produce the characteristic appearance of albinos. Partial albinism most frequently affects the hair, genitalia, nipple, and face. The influence of heredity in bringing about the defective development seems questionable, since albinos, although there may be several in one family, are usually the children of ordinarily pigmented individuals, or, indeed, they may be the offspring of negro parents.

Abnormalities of the cutis vera and subcutaneous tissue include the congenital variations affecting the connective tissue of the skin. At times the latter may be of such unnatural density, toughness, and rigidity (*scleroderma congenita*) as to appear tightly drawn, leathery, and unwrinkled. In other cases the same layers may be so loose (*cutis laxa*) that the integument and subcutaneous tissue hang in pendulous folds and admit of unusual extension, recalling the "elastic skin man" of the exhibitions.

In conditions favoring stasis of the venous and lymphatic currents of the subcutaneous tissue, the latter may become the seat of great hypertrophy with enormous distention and infiltration (*elephantiasis congenita*). This condition may affect the integument to such degree that the parts involved lose all resemblance to their normal form. Elephantiasis is frequently observed in cardiac monsters in which the subcutaneous tissue sometimes becomes a misshapen oedematous mass, owing to the infiltration of the areolar tissue.

Dermoid cysts of the skin are not infrequent. These congenital formations are to be distinguished from the true dermoid growths which represent the abortive de-

velopment of all three germ layers. The ones here under consideration result from abnormal inclusion and isolation of epithelial tissues, which later develop in unusual situations cystic tumors which may attain large size and contain within their walls epidermic deviations, such as hair follicles and sebaceous glands. In accordance with their origin, the favorite seats of such growths are locations in which fusion of the epidermis normally takes place, as along the lines of closure of visceral furrows.

Anomalies of the epidermal derivatives include unusual development of the hair and of the glands.

Congenital abnormalities of the hair may be excessive (*hypertrichosis*) or deficient (*hypotrichosis*). The former condition, in which an unnatural profusion occurs in regions ordinarily covered only by fine down, is regarded by Unna and by Brandt¹⁴⁰ as essentially a persistence of the lanugo, or foetal hair, to the exclusion of the secondary, which commonly later appears. Regarded in this light, hypertrichosis is in fact an arrested development.

The excessive development of hair may be limited (*hypertrichosis localis*), as when confined to some small area, as part of the face in "bearded women," or in individuals possessing hairy naevi or warts; or it may include large tracts or even the entire surface of the body (*hypertrichosis universalis*), as seen in the "wild men" of the museums. In 1883 "Krao," a girl of seven, a native of Indo-China, was exhibited as "Darwin's Missing Link." She was covered with black hair, had prognathic type of face, and possessed extraordinary prehensile powers of lips and feet. Sometimes the hands and feet are uninvolved, as in the case of "Jo-Jo," celebrated as the "Dog-faced Boy." The excessive development of hair commonly takes place after birth, although heredity undoubtedly plays an important rôle in establishing the excessive anlage resulting in this abnormality. Crawford and Yule described a family of Burmah, of which representatives of three generations—father, daughter, and granddaughter—were nearly covered with hair.

Defective development of the hair (*hypotrichosis*) is comparatively infrequent, persistent complete congenital alopecia being decidedly rare. These defects are often associated with deficient formation of the teeth and, sometimes, the nails. A number of instances have been recorded of individuals who have been hairless from birth, even the lanugo having been apparently wanting, although probably partially developed. Heredity here also seems to be an important factor, since in some families congenital alopecia has been noted in successive generations. Congenital glandular anomalies of the skin affect chiefly the sebaceous glands, which may be atrophic or absent, as sometimes seen in keratomous conditions of the epidermis. The cystic growths connected with the sebaceous glands (*atheroma*), formerly regarded as always due to accumulation of secretion, sometimes are probably referable to remains of a congenitally displaced epithelial anlage as a point of origin (Chiari¹⁴¹). An excellent presentation of the congenital skin, as well as other diseases affecting the fetus will be found in Ballantyne's recent work,¹⁴² to which the reader is referred.

Polymastia, the condition of having more than two breasts, is the most important congenital glandular abnormality connected with the integument, since the mammae in principle are only modified sebaceous glands. Congenital absence of one or both breasts is very rare. Numerical redundancy, on the contrary, is common, supernumerary organs being frequently encountered in both sexes. A distinction, however, must be made between the presence of merely supernumerary nipples (*hyperthelia*) and of additional true milk glands (*hypermastia*), the latter condition being much rarer than the former. The examinations recorded by Bardeleben,¹⁴³ including over eleven thousand German soldiers, gave the astonishing results that supernumerary nipples occurred in fourteen per cent., or once in every seven individuals. A percentage so high is probably obtainable only by regarding doubtful cases of pigmented spots as of sufficient significance to be included. Supernu-

merary nipples seem to be more frequently developed in men than in women; accessory true glands, however, are much more often found in female subjects than in male. The most common situations for the additional nipples are below the position of the normal glands, somewhat nearer the midline; they may occur on one or on both sides, more often, however, on the left side. Other locations of the aberrant nipples generally follow a line passing from the axilla toward the groin; exceptionally they may be upon the back or outer surface of the thigh.

The existence of supernumerary mammary glands is much rarer than of the nipples and is more frequent in women than in men. The positions of such mammary glands in women include the back, shoulder, axilla, thorax, abdomen, groin, labium majus, and thigh. Trustworthy records show that such organs secreting milk have been located in the axilla, on the chest and the abdomen, in the groin, and on the external surface of the thigh. Hirst¹⁴⁴ refers to a negress of nineteen years who possessed nine mammary glands, five on one side and four on the other. Two of the accessory organs were small and did not secrete; all the others gave milk in large quantities.

The investigations of O. Schultze¹⁴⁵ have shown that the mammary glands are developed along the *milk lines*, ridges of thickened epithelium extending from the dorsal surface, in the vicinity of the fore limbs, ventrally to end in the inguinal region. These observations at once point out the correspondence between the location of the majority of supernumerary nipples and milk glands and the normal formation tracts, the excessive mammary of the human subject marking unusual development of possible analges which ordinarily remain quiescent. The influence of heredity seems to be uncertain, although in a number of reported cases both mother and daughter presented polymastia, although the location of the supernumerary mammary was not identical.

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LITERATURE.

The enormous and ever-increasing volume of the literature relating to malformations forbids any attempt to present systematically in these pages even a synopsis of teratological bibliography. Fortunately this is also unnecessary, since those especially interested will find a practically complete bibliography since 1886 in the Anatomischer Anzeiger and the Centralblatt für allgemeine Pathologie, as well as in Schwalbe's "Jahresberichte der Anatomie u. Entwicklungsgeschichte." The "Reports on Teratological Literature" by Windle, which appear from time to time in the Journal of Anatomy and Physiology, are also useful. A valuable general bibliography, including the most important papers, is appended to Marchand's article "Missbildungen" in Eulenburger's "Real-Encyclopädie." For the older literature the reader can consult with advantage Förster, "Die Missbildungen des Menschen," 1861, and Ahlfeld, "Die Missbildungen des Menschen," 1880. A comprehensive list of journal references and monographs, appearing between 1880 and 1901, will be found in Hirst and Piersol's "Human Monstrosities."

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TERATOMA.—A teratoma is a tumor-like growth characterized essentially by the fact that the tissue-formation of which it is composed does not occur normally in the affected region, or at least not during that period of bodily development in which it presents itself. Such a growth may consist of a single tissue, or may be represented by a cyst (*simple teratoid tumor* or *cyst*); but more frequently it is made of several tissues arising from more than one of the germ layers (*mixed tumor*, "*Mischgeschwulst*"). To the more complicated forms the term *teratoma* is often applied in a more narrow sense, while those highly complex growths which contain tissues derived from all three of the germ layers are known as *embryoid tumors* or *embryomata*. Teratomata composed of tissue-formations which are out of place in the region where found are known as *heterotopous*; those composed of tissue occurring in a given region at a time when it should not normally be found there are known as *heterochronous*. The tissue-formation composing the teratoma takes its origin either from the anlage of the individual affected (*monogerminal, endogenous, or autochthonous teratoma*), or from the anlage of a second individual (*bigerminal or ectogenous teratoma, fetus in fetu*). Further, either a sarcomatous or a carcinomatous proliferation may develop within a teratoma, and the growth thereby assume the characteristics of a malignant tumor (*malignant teratoma*).

According to their structure, teratomata may be divided into the following varieties:

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|--------------------------|----------------------------|---|---------------------------|------------|--|
| Teratomata. | 1. Simple teratoid tumors. | { | Ectodermal. | { Dermoid. | |
| | | | 2. Simple teratoid cysts. | { | Mesodermal. |
| | | | | | Entodermal. |
| | | | | | 3. Complex teratomata and teratoid cysts (embryoid tumors and embryomata). |
| 4. Malignant teratomata. | | | | | |

I. SIMPLE TERATOID TUMORS.—In this class are placed both heterotopous and heterochronous tissue-formations of simple structure, consisting of but a single variety of tissue, or at most of but a few forms of tissue. For the greater part they consist of heterotopous tissue-proliferations which have their origin in misplaced or persistent fetal anlage. They may occur in any part of the body, but are found most frequently in certain regions. The most common tumors of this class are the hypernephroma (*kidney, kidney region, liver, etc.*), rhabdomyoma or rhabdomyosarcoma (*kidney, testicles, vas deferens, heart, va-*