

was in excess of 6 years. Additional features of this syndrome are significant motor and mental retardation, and an unusual facies including coarse eyebrows, prominent eyes, and low, upturned nose. Respiratory infections have been a major clinical problem. One patient died at 20 months of age from a hemorrhagic pneumonia and his autopsy did not reveal any cellular explanation for his condition. The other patient is having severe respiratory infections at 11 months of age. Both patients have been extensively investigated for the presence of endocrine or storage disorders, and no cellular or biochemical abnormalities have been detected. Although the etiology of this disorder remains unknown, the mechanisms underlying this syndrome may enhance our understanding of control mechanisms beside androgen, estrogen, or thyroid hormone which affect the rate of skeletal maturation.

Presented at the Western Society for Pediatric Research meetings. This material is of sufficient importance to be presented to a national audience.

- 23 *Genesis and Significance of Palmar and Digital Creases.* GREGORY A. POPICH and DAVID W. SMITH. Dept. of Ped., Univ. of Washington Sch. of Med., Seattle, Wash.

Our studies of skin creases in the early developing hand and in a variety of malformed hands show that these creases, which develop between 7 and 14 weeks of fetal life, are secondary to the early flexion planes in the developing hand. Thus with lack of development or flexion of a digital joint no underlying digital crease was found; and with lack of formation or oppositional function of the thumb there was no thenar palmar crease. The normal distal palmar sloping crease was found to be related to the sloping flexional plane of the 3rd, 4th, and 5th metacarpal-phalangeal joints and the usual mid-palmar crease represents the plane of skin folding between the sloping distal crease and the thenar crease. The occurrence of a single horizontal palmar crease, the simian crease, was found by roentgenographic studies to be related to a diminished slope of the 3rd, 4th, and 5th metacarpal-phalangeal joints such that only a single early horizontal plane of flexion existed in the skin of the upper palm.

Thus hand creases provide a record of the early flexional planes of function in the developing hand and any crease abnormality should always be interpreted as an indication of a more primary defect in form and/or function of the early developing hand.

- 24 *Severe Pyridoxine Deficiency: A Model for Ataxia Telangiectasia.* THOMAS E. NELSON, STARKEY D. DAVIS and THOMAS H. SHEPARD, Univ. of Wash. Sch. of Med., Dept. of Ped., Seattle, Wash.

The association of neural, vascular, and immunologic abnormalities in ataxia telangiectasia is unexplained. We studied the teratogenic effects of B₆ as an experimental model because it is associated with (1) neonatal convulsions and (2) impaired immunologic response in rats. Pregnant rats were given a B₆-deficient diet and 4-desoxypyridoxine, an inhibitor of B₆. The control group had the same plus excess pyridoxine. The fetuses were examined at 20 days. Treated rats developed clinical signs of B₆ deficiency and gained less weight than the controls. Mean weight of 108 B₆-deficient fetuses was 1.63 g; mean crown rump length was 28.3 mm. The 61 control fetuses had a mean weight of 3.24 g and a mean length of 37.0 mm; no anomalies were found in the control group. Of the treated group, 48%

had digital defects, 20% had cleft palates, 12% had omphaloceles, 8% had micrognathia, and 6% had exencephaly.

Organ weights (as g % formalin fixed body weight)

Fetus	Kidney	Thymus	Spleen
Treated (21)	0.88	0.19	0.03
Control (25)	0.85	0.24	0.14

Though the thymus and spleen are both significantly smaller ($p < 0.001$) in the treated group, only the spleen is strikingly hypoplastic. We anticipate that intrauterine B₆ deficiency will cause a lifelong immunologic defect.

- 25 *The Production of Multinucleated Lymphocytes by Cytochalasin B. An Electron Microscopic Study.* G. F. SMITH, P. O'HARA and M. A. C. RIDLER, Dept. of Ped., Loyola Univ. Stritch Sch. of Med., Maywood, Ill., and Kennedy-Galton Lab., St. Albans, England.

The cytochalasins are newly derived from moulds and are useful in studying nuclear division. Multinucleated fibroblasts were produced by CARTER [Nature 213: 261, 1967] and lymphocytes by us [Nature 216: 1134, 1967]. Lymphocytes with up to eight nuclei were seen. Progressive nuclear addition [1, 2, 3, etc.] was found instead of a doubling of the nuclei (1, 2, 4, etc.). The term 'pseudomitosis' was proposed for the process of synchronous mitosis, but which results in the division of only one nuclei.

Our observations, using the light microscope shows the nuclei of binucleated cells to be similar in size and shape; however, in cells with more than two nuclei there is marked variation in size and shape of each nucleus. What might have appeared, in many instances, as separate nuclei are seen by electron microscopy to be lobes of a rather large and irregular nucleus. In other instances, small nuclear bridges connected what might appear as separate nuclei. Variation exists in nuclear size from rather large nuclei to very small micronuclei. Serial sectioning of cell nuclei verified the morphological variability seen in single sections. Spindle fibers and kinetochore attachment to the chromosomes revealed no unusual features. Electron microscopy showed synchronous division of nuclei. Golgi apparatus remained as a single large unit. There was a doubling of centrioles in binucleated cells. What appears as separate nuclei under the light microscope were connected by nuclear bridges. The finding suggests that as nuclei accumulate that the spindle bridge is unable to accurately divide the large numbers of chromosomes present.

- 26 *Phocomelia With Mandibular Hypoplasia: A New Syndrome.* SALMA REGINA ASSEMANI, TADASHI KAJI and LYTT I. GARDNER, State Univ. of New York, Upstate Med. Center, Dept. of Ped., Syracuse, NY.

We have observed the association of non-thalidomide phocomelia and hypoplasia of the mandible (Pierre-Robin syndrome) in two unrelated infants. Baby boy C was born at term after a normal gestation. The mother was 29 and denied either illness or medication during pregnancy. Five months postpartum she had no glucosuria. The baby showed multiple deformities: both hands were hypoplastic with no visibly separate fingers. The right hand was more primitive than