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We hope that making available the relevant information on Pachyonychia Congenita will be a means of furthering research to find effective therapies and a cure for PC.
of methyl parathion insecticide in the form of snuff produced similar lesions. To our knowledge, erythema multiforme produced by inhalations of insecticide, particularly Dalf, has not yet been reported.

R. K. BHARGAVA
VIRENDRA SINGH
VINAY SONI
Jaipur-4 (Raj), India

Pachyonychia Congenita With Cardiac Involvement

To the Editor.—A case of pachyonchyia congenita with cardiac involvement has been recorded and reported. A 9-year-old girl had all the characteristic features of pachyonchyia congenita. In addition, there was clinical and roentgenographic evidence of cardiacomegaly, with abnormal ECG and angiographic studies. She also had a pigmented nevus in the left iris, spinifibida occulta L5, radioumeri 12th rib, sparse kinky hair over the scalp, and curious teeth.

Report of a case.—A 9-year-old girl born to consanguineous parents was brought to the Dermatology Department on Aug 18, 1974. When her history was taken, it was noted that approximately three months after birth blisters in frictional areas started to develop. At the age of 7 months, horsy follicular lesions mostly over the extremities developed. Nail dystrophy in the form of thickening and lifting up of the nail plates started at the age of 1 year. When the child started walking, blisters developed on the soles and lead to painful peeling off of the skin. At the age of 6 years, curious teeth developed as did sparse kinky hair over the scalp. History was suggestive of cyanotic spells with crying that started at the age of 2 years and persisted until the age of 5 years.

On examination she was moderately nourished and slightly anemic. Dermatological examination showed sparse kinky hair over the scalp, keratoderma on the palms and soles, follicular papular lesions over the extremities, verrucose plaques over the elbows, knees, and gluteal folds, large ruptured bullae with exfoliation in both soles, and leukokeratosis in the buccal mucosa. All the nail plates were thickened with subungual hyperkeratosis and lifting up of the nail plates distally. Systemic examination showed clinical and roentgenographic evidence of cardiacomegaly; the pulmonic second sound was loud and split; her pulse rate was 108 beats per minute; and blood pressure was 100/80 mm Hg. Fluorescence revealed cardiomagi. The C/T ratio was 60:40. Visible pulsations and minimal enlargement of both ventricular chambers were present. Electrocardiogram showed normal sinus rhythm, left anterior fascicular block, and nondiagnostic T wave changes anterolaterally. Though mere T wave with ST-Segment changes can happen in any primary myocardial disease, the Q-S pattern in V5 and V6 was more in favor of a specific anamnestic origin of coronary artery from pulmonary artery.

The angiography report by Dr T.J. Cheriyan was as follows:

Catheter in the pulmonary artery in the AP view shows normal pulmonary artery and its branches. There is an obstruction of the left atrium and left ventricle opacified normally. The left ventricular contraction appears to be sluggish but there was no evidence of thinning of the ventricular musculature. Aortic root injection in AP view shows both right and left coronary arteries filling normally and the right coronary artery appears to be the dominant artery. The left ventricular injection in the RAO shows a dilated left ventricle with a number of premature ventricular contractions but in the later phase ventricular contractions appear to be sluggish. The overall impression was suggestive of myocarditis.

Correction of the anemia did not improve her cardiac condition.

Comment.—This case had all the characteristic features of pachyonchyia congenita as described in the literature. In addition, this child had a cardiac abnormality along with an abnormality of the left iris that to our knowledge have not been reported in the literature so far. It is worthwhile in the future to look for a cardiac abnormality in cases of pachyonchyia congenita to decide whether this association we have reported is merely fortuitous or whether a cardiac anomaly might also occur in a full-figured case of pachyonchyia congenita.

S. PREMALATHA, MD, DD
A. S. THAMBIAH, FRCP, FAMS
Madras, India

Dr T. J. Cheriyan, physician, did the investigations and the report on the cardiac condition.

Toxic Epidermal Necrosis in a Patient With Ovarian Carcinoma

To the Editor.—Toxic epidermal necrosis is recognized with increasing frequency in association with an enlarging variety of other diseases and is apparently precipitated by a very wide spectrum of triggering mechanisms. Rarer but important associations have also been reported and we would like to report another example in this category of toxic epidermal necrosis occurring in association with a systemic neoplasm.

Report of a Case.—A 66-year-old housewife had a hysterectomy for fibroids 16 years previously and had been active and well until recently when indigestion and water brash developed and she began to lose weight. Her epigastric pain was related to eating food and the pain radiated to the back and was adequately relieved by antacids. On examination, the only abnormal physical sign was an aortic systolic grade 2 murmur.

Laboratory studies on admission revealed an ESR of 104 mm/hr, a hemoglobin value 11.4 gm/100 ml, an erythrocyte count of 3.8 x 1012/liter, and a leucocyte count of 6,000/cm3 with typical differential distribution. The plasma urea value was 84 mg/100 ml. Plasma potassium was persistently low at a value of 2.8 mEq/liter. Liver function tests results showed strongly positive thymol flocculation and thymol turbidity at 9 MacLagen units. The urinary protein level was 30 mg/liter of urine. No Bence-Jones protein was detected. Immunoglobulin IgA and IgM values were normal, but the IgG level was raised at 21.40 gm/liter. Other investigations reported were normal were x-ray films of the chest and abdomen, barium meal examination of the esophagus, stomach, and duodenum and findings from biopsy of the liver and of the pectoral temporal region. Results from the bacteriological examination of the skin, nose, mouth, and sputum were all reported to be normal apart from the usual commensals. There were no lpus erythematosus cells in the blood and results from immunofluorescint tests for antinuclear, mitochondrial, and smooth muscle antibodies were all normal. Her protein-bound iodine value was within normal range at 5.6 mg/100 ml.

The patient continued to receive antacids and was about to be discharged home to await a lymphangiogram when an itchy vesicular eruption rapidly developed on the legs and trunk. Swabs taken from these sites for bacteriological culture were reported negative for Staphylococcus aureus and other pathogenic organisms. After several days, the skin rash developed into a blistering skin eruption that showed grouped tense clear vesicles on erythematous bases scattered over the trunk and elbows.

A provisional diagnosis of bullous pemphigoid or dermatitis herpetiformis was made at this time. A biopsy specimen was taken for histological examination and serum for immunofluorescence tests. Dapsone 50 mg twice daily was prescribed. Six days later, however, the patient's condition rapidly deteriorated. In the words of her own physician, "Her skin had come off overnight" and she was found to have the characteristic eruption of toxic epidermal necrosis. There was trunk and limbs were involved with widespread areas of erythema on which were scattered clear vesicles and bullae. Certain denuded friction sites looked like severe scalds. The patient was given prednisolone phosphate 80 mg daily and during the first week of this treatment