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Cutis Laxa with Growth and Developmental Delay

To the editor:

Congenital cutis laxa is a rare inherited disorder of connective tissue manifested by loose, hanging skin. Cutis laxa may be inherited (autosomal dominant, autosomal recessive) or acquired. As opposed to the recessive form of cutis laxa, the dominant form has been reported to be free of pulmonary and other internal manifestation.[1] Skin fibroblast cultures from cutis laxa patients exhibit reduced elastin. The genetic map locus of autosomal dominant form is 7q11.2. Tassabehji et al[2] have described patients with autosomal dominant cutis laxa and mutations in elastin gene.

Congenital cutis laxa with growth and developmental delay has recently been defined as a distinct entity of autosomal recessive inheritance.

In this report, we described a patient with cutis laxa and growth retardation.

This 1.5-year-old girl was admitted to our hospital for bilateral inguinal hernias and lax skin. The patient was the third child born by spontaneous vaginal delivery at term to a 28-year-old G3P4 mother and 30-year-old father who were both healthy. Birth weight and length were not known. The parents were third-degree relatives. The paternal uncle also had 2 daughters with the same disease.

On physical examination weight and height were 9 kg and 74 cm, respectively, both of them being below the 3rd percentile. Examination revealed bilateral inguinal hernias. There was no hyperextensibility of the joints (Figure 1).

The skin of the patient was extremely redundant, the folds being soft, doughy, and inelastic. These changes were most marked on the face, arms, legs, neck, and abdomen. She had long philtrum and wide nasal bridge.

Cardiac examination revealed apical midsystolic click and 2/6 systolic murmur. Radiographic examination of the chest and gastrointestinal system and abdominal ultrasonography appeared normal. Echocardiography revealed mitral valve prolapse. A skin biopsy was obtained and processed for light microscopy. It revealed a decrease in the number of the elastic fibers. Chromosomal analysis yielded normal findings.

Under the title "congenital cutis laxa with growth retardation and developmental delay," 27 patients have been reported. The condition has been described as a distinct entity of autosomal recessive inheritance.[3] It is recognizable at birth owing to its typical skin findings. Even at birth, there are multiple folds of loose redundant skin over the trunk and ex-

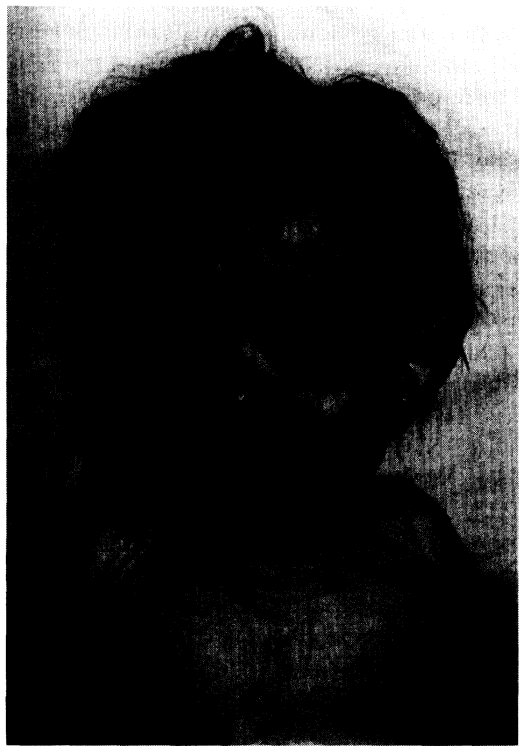

Figure 1. Photograph of the patient with marked laxity of the skin.

tremities. Involvement of the face with sagging skin gives the patients a senile appearence.[4] This problem increases with age. Ectropion and blepharochalasis of the lids may also be noted. Congenital dislocation of the hip, diverticula of bladder and gastrointestinal system, hydronephrosis, prenatal and postnatal growth retardation, wide or delayed closure of fontanelle, reversed eyebrows, bilateral inguinal hernias, long philtrum, and wide nasal bridge, have all been reported in this syndrome.[5]

Because it is a rare and interesting syndrome, we wanted to present this patient.

C. Karakurt
T. Sipahi
*S. Ceylaner**
F. Şenocak
S. Karademir
M. Becer
*Dr. Sami Ulus Children's Hospital and *Zekai Tahir Burak Maternity Hospital, Department of Prenatal Diagnosis and Genetics, Ankara, Turkey*

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Bone Mineral Density in Adolescents

To the editor:

In the article by Cromer and Harel,[1] the attainment of an optimal peak bone density was reported as the most important factor for preventing adolescent osteoporosis. The authors suggested that during the evaluation of bone mineral density dietary calcium intake, physical activity, and sex hormone therapy, especially subdermal implants or combined oral contraceptives, should be considered.

Prevention of disease is an important aim of medicine. Although a significant geriatric problem, osteoporosis may be prevented in childhood and adolescence. In our country, postmenopausal osteoporosis is common, and male osteoporosis has also been reported in men 50–70 years of age.[2] Because adolescence is a critical period for the deviopment of osteoporosis, we planned to study the bone mineral density in adolescents. Fifty-two adolescents, 26 girls and 26 boys who attended Social Security Ankara Children's Hospital, were included in this study. Exclusion criteria were endocrine, renal, and gastrointestinal disease and use of drugs that might affect calcium and/or vitamin D metabolism. All adolescents had similar socioeconomic and cultural conditions, and none of them had a history of regular physical activity. Bone mineral density was measured by dual-energy x-ray absorpsiometry. Twelve girls and 13 boys were osteopenic [–1.0–2.5 standard deviation (SD)], whereas 1 girl and 4 boys showed overt osteoporosis [over –2.5 SD].

We have started a training program for parents of children and adolescents at our hospital with brochures and booklets. We would like to emphasize the importance of educating parents and adolescents about osteoporosis.

Fatma Nur Cakmak, MD
Didem Aliefendioglu, MD
Hulya Erdem Ayas, MD
Ergun Cetinkaya, MD
Vildan Kosan, MD
*Levent Kuscu**
*Social Security Children's Hospital and *Duzen Laboratories, Ankara, Turkey*

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