

ADRENOLEUKODYSTROPHY: INCREASED PLASMA CONTENT OF SATURATED VERY LONG-CHAIN FATTY-ACIDS

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With a new method we measured the saturated very long chain fatty acids in the plasma of adrenoleukodystrophy (ALD) hemizygotes, ALD heterozygotes, and controls. ALD hemizygotes showed increased levels of hexacosanoate (C26 fatty acid) which represented $0.081 \pm 0.0066\%$ (SEM) of total fatty acids, compared to $0.015 \pm 0.0032\%$ in the controls. C25, C24, and C23 fatty acids were also increased, but the C22 and C20 fatty acids were normal. C26 levels were also increased in most ALD heterozygotes, with a mean level $0.057 \pm 0.0063\%$ of total fatty acids. The technique can be used for diagnosis and carrier identification, and in the evaluation of therapy.

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Comment from Jonathan W. Mink, MD, PhD, FAAN, Associate Editor: This was the original report of elevated very long chain fatty acids (VLCFA) in the serum of hemizygous males with x-linked adrenoleukodystrophy (ALD) and in heterozygous female carriers. Measurement of serum VLCFA remains the primary mode for definitive laboratory diagnosis of this x-linked ALD.

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