

MRI : 1

1

Burlina

10

(1.5 T Signa Horizon;

21 가 (1, 2).

General Electric Medical System, Milwaukee, WI, U.S.A.)

T1

, T2

(Fig. 1A - C).

T1

, T2

(3).

1

(Fig. 1D, E).

14 가

가

1

가

가 가

(Fig. 2).

3

4

1

가 3356/ul serum lac -
tate가 8.73 mmol/l, WBC count가 23,900/ml 가
, platelet count, PT aPTT

2

가

가

acylcarnitine C4 (butyrylcarnitine and isobu-
tyrylcarnitine) C5 (2 - methylbutyrylcarnitine and iso-
valerylcarnitine) 가

short, medium, and long chain acyl CoA dehydro-
genase electron transfer flavoprotein and ETF -
ubiquinone oxidoreductase

Burlina 1991
branched - chain acyl - CoA dehydrogenase

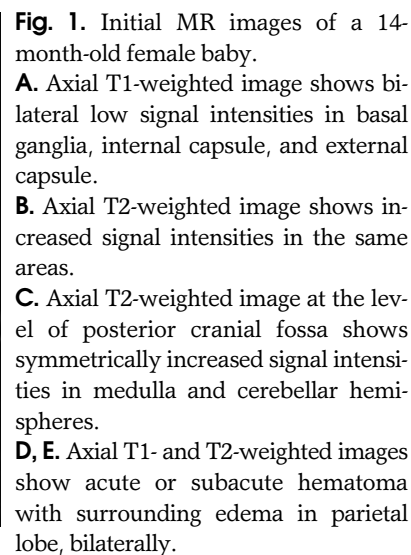
branched - chain acyl - CoA dehydrogenase

, 1994
2 - methyl

(2).

•

(3).



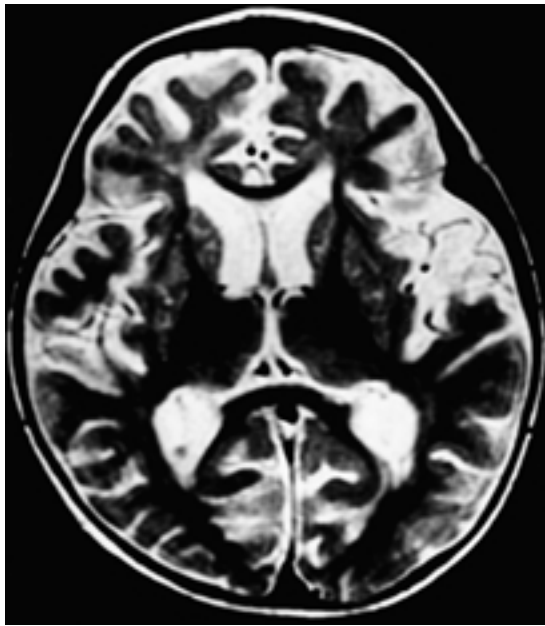


Fig. 2. Follow-up T2-weighted image after 1 month. MR image shows progression of diffuse brain atrophy and shrinkage of the caudate and lentiform nucleus with some heterogenous signal intensities.

(7).

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(6),

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MRI Finding of Ethylmalonic Encephalopathy: Case Report¹

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Ethylmalonic encephalopathy is a rare syndrom characterized by developmental delay, acrocyanosis, petechiae, chronic diarrhea, and ethylmalonic, lactic, and methylsuccinic aciduria. We report the MRI finding of ethylmalonic encephalopathy including previously unreported intracranial hematoma.

Index words : Brain, diseases
Brain, metabolism
Brain, MR
Brain, hemorrhage
Children, central nervous system

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