

Alloisoleucine levels in 3 patients with classical Maple Syrup Urine Disease after Orthotopic Liver transplantation

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BACKGROUND

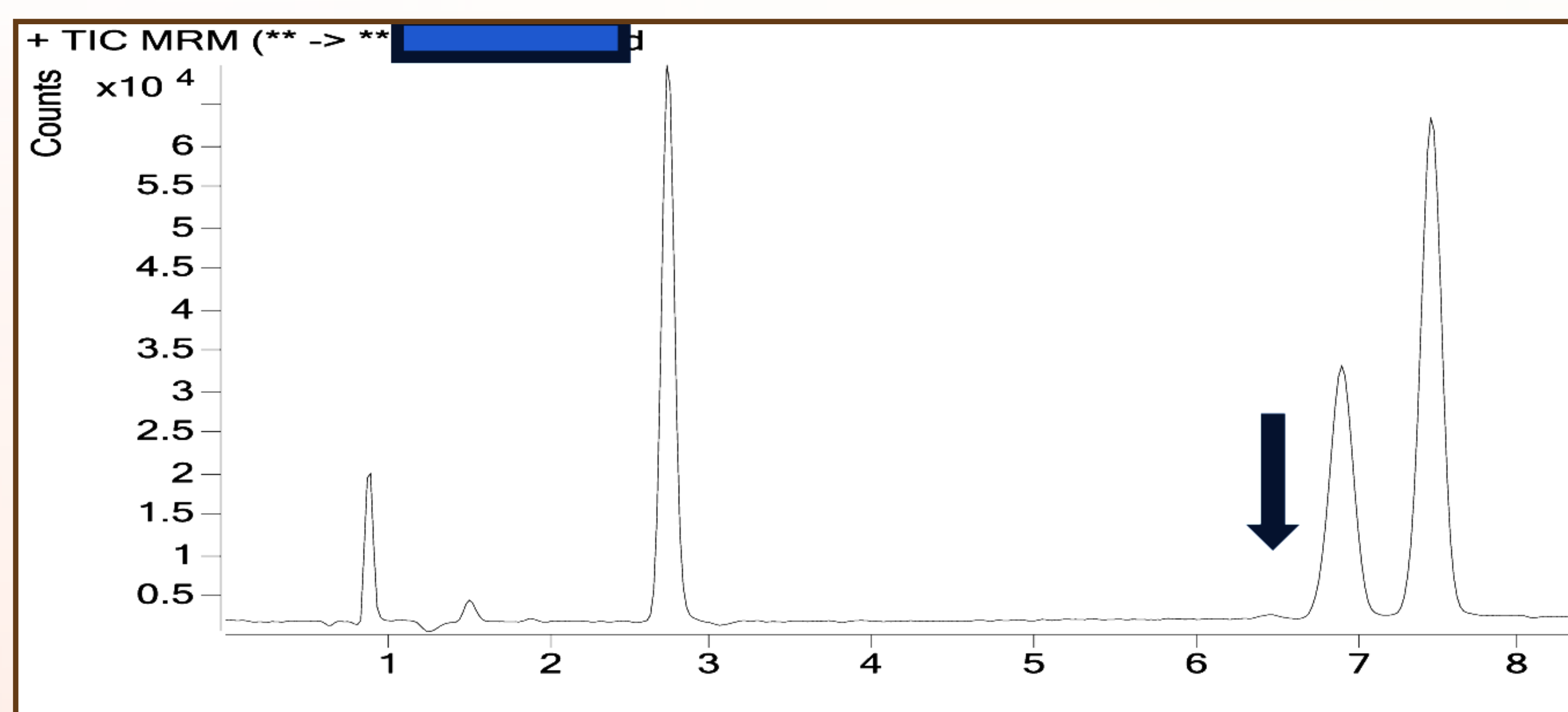
MSUD is an autosomal recessively inherited deficiency of BCKDH which catalyzes the metabolism of BCAAs. Alloisoleucine derived from isoleucine by transamination of 2-keto-3-methylvalerate is a specific and sensitive marker for MSUD. We herewith present the data on effect of liver transplantation (OLT) on levels of alloisoleucine in 3 patients with classical MSUD.

MATERIALS AND METHODS

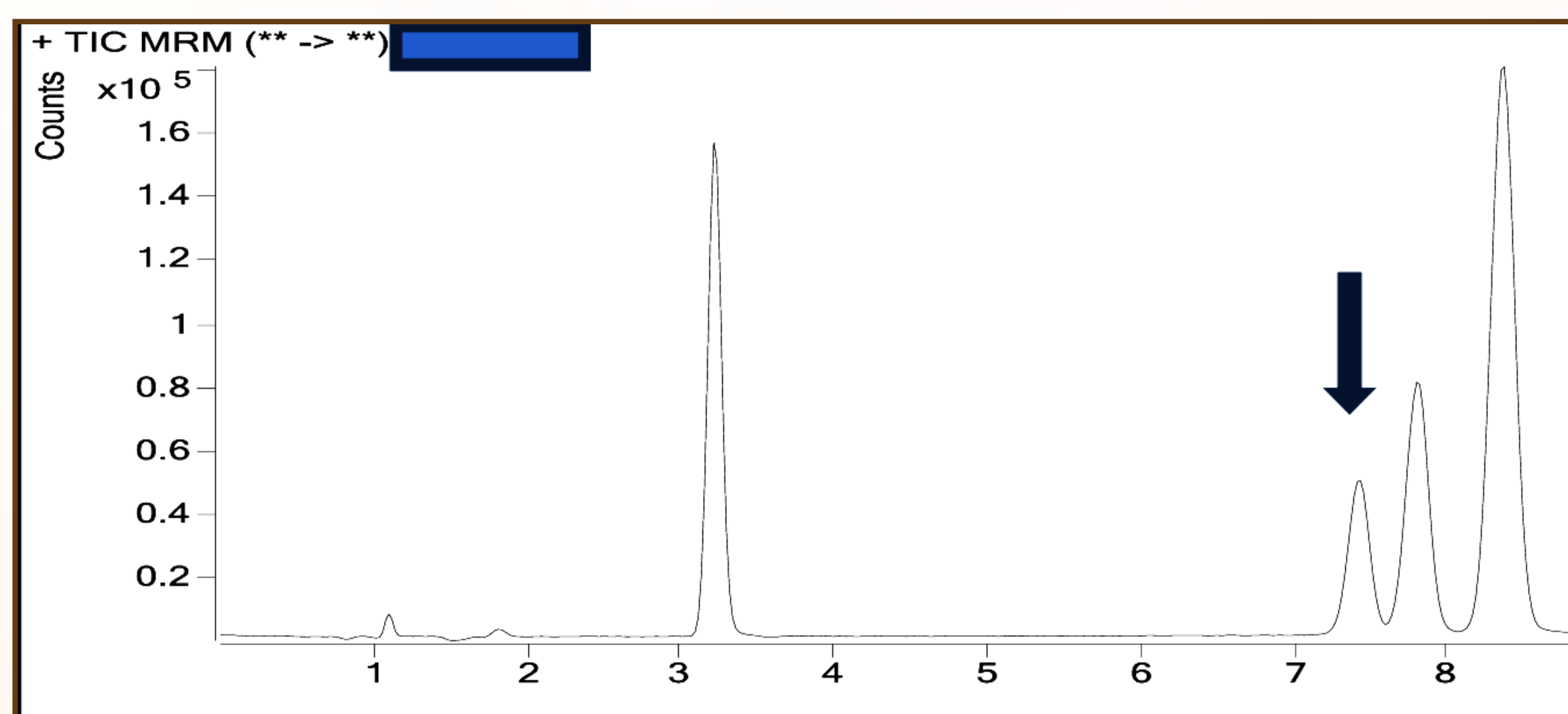
OLT was performed at ages 23months, 7months and 5yrs in 3 patients. Aminoacids including BCAA - Leu, Ile, Val and alloisoleucine were monitored on a regular basis. Two of these patients received unrestricted diet post OLT and one succumbed to sepsis. Data on alloisoleucine levels in these patients were analyzed retrospectively to determine the usefulness in monitoring.

RESULTS

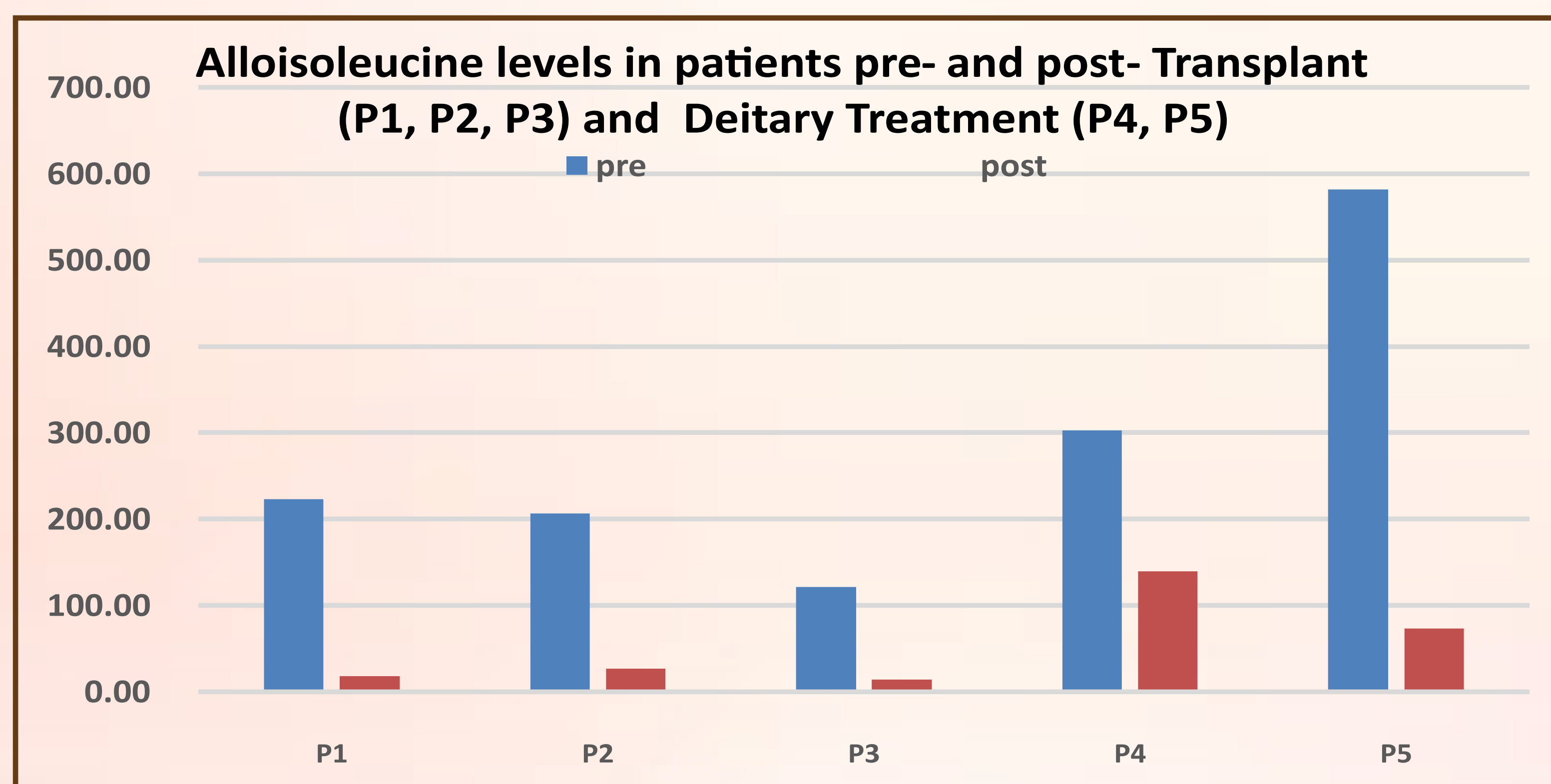
Groups	N=	Alloisoleucine Levels (µm/L)
Non- MSUD Controls	37	0.55 ± 1.21
Classical MSUD- Untreated	40	303.30 ± 207.99 (Median : 245.42)
Classical MSUD- On Dietary Treatment	8	242.02 ± 149.28
Classical MSUD- After OLT	3	31.12± 18.57



Non- MSUD patient. Alloisoleucine is undetectable



MSUD patient shows elevated Alloisoleucine



Patient	% reduction in Allo
Patient 1 (transplanted)	92.01%
Patient 2 (transplanted)	91.55%
Patient 3 (transplanted)	88.02%
Patient 4 (Dietary restriction)	53.93%
Patient 5 (Dietary restriction)	87.42%
Average reduction-Transplant (n=3)	90.53%
Average reduction -Dietary treatment (n=8)	38.32%

DISCUSSION

- Classical MSUD subjects had high alloisoleucine
- It was undetectable in most normal subjects.
- Patients on MSUD diet showed mild reduction of alloisoleucine levels (Average reduction: 38.32%)
- Prior to OLT the BCAAs and alloisoleucine levels were higher in all 3 patients despite them being on MSUD diet.
- After OLT they received unrestricted diet with normalization of Leu, Ile and Val within 1-2 weeks of OLT.
- Alloisoleucine levels drastically reduced even though they were mildly elevated as compared to normal controls. (Average reduction: 90.53%)

CONCLUSION

We highlight the utility of Alloisoleucine monitoring in MSUD patients after OLT as it is rapid and can identify control and success of management after OLT. OLT shows much better reduction of alloisoleucine levels despite increase in natural protein intake as compared to those treated with dietary restriction and special formulas.

References: 1. Significance of L-Alloisoleucine in plasma for diagnosis of MSUD. Schadewaldt P, et al. Clinical Chemistry. 1999;45:10
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3. Elective liver transplantation for the treatment of classical MSUD. K A Strauss, et al. American Journal of transplantation 2006; 6: 557-564.