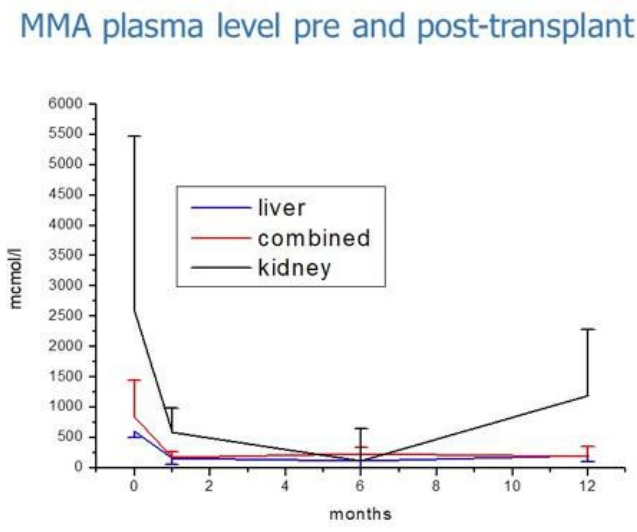
acid, leading to recurrent metabolic decompensation episodes and to chronic organ injury. Recently, there has been a growing interest in organ transplantation approaches: kidney transplantation (KTx) corrects renal failure but only partially restores the enzyme activity which is better obtained by liver (LTx) or combined liver/kidney transplant (LKTx). A consensus on the optimal transplant strategy is lacking. Our aim was to compare in a large multicenter study the results of different transplant approaches in patients affected by MMA.

Material and methods: Within centers of the Cooperative European Paediatric Renal Transplant Initiative (CERTAIN) Registry and additional centers both in the US and in Europe (thanks to the joint effort of ESPN and MidWest consortium) we collected retrospective clinical and biochemical outcome data from transplanted patients with MMA from the year 2000 to present.

Results: So far, 58 patients were registered. Currently, data are available for 34 patients: 19 with KTx, 7 with LTx and 8 with LKTx. Median age (years) at transplant was 13.6 for KTx, 2.5 for LTx and 8.2 for LKTx. Median pre-transplant plasma MMA was higher in patients with KTx (2572 µmol/L) than in LTx (599 µmol/L) or LKTx (836 µmol/L). Post-transplant MMA was evaluated at 1, 6 and 12 months (Figure). At 1 and 6 months, all patients had a clear reduction of plasma MMA, whereas at 12 months only KTx patients showed a negative trend with a return plasma MMA increase. LTx and LKTx patients maintained significantly lower MMA plasma levels.



At transplantation, eGFR in LTx and LKTx was 87 and 40 ml/min/1.73 m2. KTx patients were in advanced chronic renal failure. At month 12, renal function was still normal in LTx or LKTx (median eGFR 104 and 89 ml/min/1.73 m2), while in KTx it was only 49 ml/min/1.73 m2. At last follow-up, 21% of pts with KTx and 13% with LTx or LKTx were deceased. Neurological outcome at 1 year post-transplant was stable in all patients.

Conclusion: These data show a more pronounced and persistent post-transplant reduction of plasma MMA levels following LTx or LKTx compared to KTx alone. Graft and possibly patient survival is suboptimal following isolated KTx, where impaired graft function likely leads to reduction in renal filtration of MMA and lower intrarenal enzymatic activity, both of which increase MMA plasma levels driving further decline of renal function.

401.7 | Intra-operative near-miss events during left liver lobe donation surgery: The hidden face of paediatric living-donor liver transplantation

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Introduction: Living donor liver transplantation (LDLT) was developed to alleviate post-mortem organ shortage in pediatric liver transplantation. Such program, however, should be performed under strict ethical guidelines, and optimal living donor (LD) safety. We hypothesized that serious unexpected peri-operative medical and/or surgical events during LD procedure indeed occur in a high volume LDLT center, being a major threat to donor safety.

Methods: Between July 1993 and October 2018, a total of 433 paediatric LDLT were performed at Cliniques universitaires Saint-Luc, Brussels Belgium. Each LD was carefully selected through medical history, pre-operative work-up, medical advocate and psychologist consultations, and liver US and MRI imaging. Left liver lobectomy was performed through a midline laparotomy as previous detailed (1). All LD medical records were retrospectively reviewed searching for the occurrence of near-miss events (NME). NME was defined as any potentially harmful event during peri-operative period that was identified and controlled before definitive patient harm has happened. No recognition/no control of such events constitute a potential risk for subsequent donor and recipient morbi-mortality.

Results: Paediatric recipients results were described elsewhere (1). Neither mortality nor persisting disability was encountered in our series. Morbidity consisted mainly in cut-surface bile collections, transient pleural effusions, and incisional hernias, in less than 3% each. The following 8 (1.85%) intra-operative NME were identified in LD (one case each): 1) tension pneumothorax secondary to right subclavian vein function, 2) accidental ligature-section of donor common hepatic duct, 3) unintended ligature-section of donor right hepatic artery (RHA),4) accidental vena cava clamp release resulting in a massive bleeding, 5) slipping of the left hepatic artery (LHA) Hemolock® clip, 6) full IV LD heparinization, 7) lateral injury of seg. II bile duct evidenced at intra-operatory cholangiography, 8) accidental section of LHA at liver graft hilum. NMEs management consisted in: chest drain insertion, Roux-en-Y biliary drainage, RHA

microsurgical re-anastomosis, temporary Pringle maneuver and total vena cava clamping followed by left hepatic vein stump suture, temporary clamping of common hepatic artery and LHA stump clipping, immediate protamine infusion, repair of bile duct injury and microsurgical reconstruction of LHA both at back-table. None of these NME resulted in peri-operative morbidity.

Conclusions: Despite our minimal morbidity and no mortality rates, unexpected potentially life-threatening intra-operative events were identified in this series. All NME were timely recognised and controlled with no donor or recipient definitive harm. Systematic debriefing of each NME conducted to safety refinements in our LD protocol. We suggest that LD/NME systematic record could be use as quality control tool to assess donor safety in LDLT programs.

References: 1. Gurevich M et al. Annals of Surgery 2015; 262: 1141-1149.

401.8 | The toronto experience with anonymous live liver donors: The pediatric lens

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Introduction: Most transplant centers require a biological relationship or a strong emotional bond between live liver donors and their recipients. We report a unique experience with 28 anonymous donors who donated a part of their liver to a pediatric recipient with whom they had no biological connection or prior relationship (A-LLD).

Methods: Candidates were screened to confirm excellent physical, mental, social, and financial health. Demographics and outcomes were analyzed. Validated self-reported questionnaires assessed personality traits and psychosocial impact. Qualitative interviews after donation examined motivation and experiences.

Results: Between 2005-2017, 28 anonymous live liver transplants (A-LDLTs) were allocated to pediatric recipients. Most donors had a university education, a middle-class income, and a history of altruism. 24 (86%) were women. Median age was 39 (21-59) years. In a 5-factor model of personality, donors identified most with agreeableness and conscientiousness; and least with neuroticism. Helping others, generativity, and reciprocity for past generosity were the most common motivators. 16 (60%) learned of this opportunity through local, national or social media. 5 (18%) were directed to a specific recipient. 4 (14%) came forward intending to donate to anyone in need and were allocated to a pediatric recipient due to volume restrictions. One-year recipient survival was 97%. The overall donor complication rate was 18% (5 patients) with a median hospital stay of 5 days. There were 2 (7%) major complications that resolved: neuropraxia and transient mild mania. One donor <22 years, who in retrospect did not have sufficiently strong social supports, required extended counselling. None expressed regret. 24 (86%) maintained anonymity. 1 (3%) was a previous kidney donor, while 3 (11%) subsequently donated a kidney.

Conclusion: Anonymous liver donors can successfully contribute to the donor organ pool. Social media can be used to educate communities and increase awareness about this opportunity. Anonymous donors are motivated by their values and beliefs and are usually very satisfied with their experience. Recovery is aided by strong social supports. This is the first and only report of the motivations and facilitators of A-LLD in a large cohort. With rigorous protocols and well-established partnership between donor (adult) and recipient (pediatric) centres, outcomes are excellent. A-LLD has significant potential to reduce the gap between transplant organ demand and availability.

401.9 | The multidisciplinary management and the early referral to a specialized center improve outcomes after liver transplantation in children with metabolic disease

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Introduction: Metabolic diseases (MD) represents the second most common indication for liver transplantation (LT) in children after biliary atresia (1-3). The transplant workup in MD is complex and requires a multidisciplinary management with the aim to improve the quality of life of patients (pts) who suffer from severe disease manifestations. However, indication and timing of LT in MD is not well established yet.

Methods: A retrospective analysis of all pts who underwent LT for MD was conducted. The pre-transplant workup included metabolic, nephrologist, dietary, surgical and neurological investigations. Brain Magnetic Resonance and rachicentesis were performed in urea