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Outcome analysis of fetuses with megacystis after intrauterine vesico-amniotic shunting

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Objective

To assess the spectrum of underlying pathologies, the intrauterine course and postnatal outcome of 46 fetuses with megacystis that underwent intrauterine vesico-amniotic shunting (VAS) with the Somatex® shunt in a single center.

Methods

Retrospective analysis of 46 fetuses with megacystis that underwent VAS either up to 14 + 0 weeks (early VAS), between 14 + 1 and 17 + 0 weeks (intermediate VAS) or after 17 + 0 weeks of gestation (late VAS) in a single tertiary referralcenter. Intrauterine course, underlying pathology and postnatal outcome were assessed and correlated with the underlying pathology and gestational age at first VAS.

Results

46 fetuses underwent VAS, 41 (89%) were male and 5 (11%) were female. 28 (61%) fetuses had isolated and 18 (39%) had complex megacystis with either aneuploidy (n = 1), anorectal malformations (n = 6), cloacal malformations (n = 3), congenital anomalies overlapping with VACTER association (n = 6) or Megacystis–Microcolon Intestinal–Hypoperistalsis Syndrome (MMIHS) (n = 2). The sonographic ‘keyhole sign’ significantly predicted isolated megacystis ($p < 0.001$). 7 pregnancies were terminated, 4 babies died in the neonatal period, 1 baby died at the age of 2.5 months and 34 (74%) infants survived until last follow-up. After exclusion of the terminated pregnancies, intention-to-treat survival rate was 87%. Mean follow-up period was 24 months (range 1–72). The underlying pathology was highly variable and included posterior urethral valve (46%), hypoplastic or atretic urethra (35%), MMIHS or prune belly syndrome (10%) and primary vesico-ureteral reflux (2%). In 7% no pathology could be detected postnatally. No sonographic marker was identified to predict the underlying pathology prenatally. 14 fetuses underwent early, 24 intermediate and 8 late VAS. In the early VAS subgroup, amnion infusion prior to VAS was significantly less often necessary (7%), shunt complications were significantly less common (29%) and immediate kidney replacement therapy postnatally became less often necessary (0%). In contrast, preterm delivery ≤ 32 + 0 weeks was more common (30%) and survival rate was lower (70%) after early VAS compared to intermediate or late VAS. Overall, 90% of liveborn babies had sufficient kidney function without need for kidney replacement therapy until last follow-up, and 95% had sufficient pulmonary function without need for mechanical respiratory support. 18% of babies with complex megacystis suffered from additional health restrictions due to their major concomitant malformations.

Conclusion

Our data suggest that VAS is feasible from the first trimester onward. Early intervention has the potential to preserve neonatal kidney function in the majority of cases and enables neonatal survival in up to 87% of cases. Despite successful fetal intervention, parents should be aware of the potential of mid- or long-term kidney failure and of additional health impairments due to concomitant extra-renal anomalies that cannot be excluded at time of intervention.