Severe hydramnios and hyperprostaglandin E syndrome

Moffat, R, Di Cienzo G, Visca E, Tercanli S, *Bührer C, Holzgreve W, Hoesli I
Women's University Hospital Basel
*Children's University Hospital Basel
Switzerland

Causes for severe hydramnios (pocket > 15 cm) can be identified in most cases by fetal ultrasonography, karyotyping, and search for maternal diabetes. A 40 year-old gravida III, para I was referred to our hospital at 23^{+6} weeks of gestation with bulging membranes, severe hydramnios (AFI 45, largest pocket 16), and a prominent fetal bladder. The patient's medical, obstetric and family history were unremarkable, and a series of investigations gave normal findings (fetal karyotype, anatomy, biometry, ACM velocity, TORCH serology, oGTT, and amniotic fluid AChE). Although attempts failed to confirm the suspected diagnosis of an inherited salt-losing nephropathy (normal chloride in amniotic fluid, physiologically elevated maternal aldosterone) at this stage, a trial of indomethacin therapy (3 x 25 mg/d) was started, with serial evaluations of the ductus arteriosus. A single amnioreduction was performed at 30 weeks (1200 ml), followed by increased indomethacin dosage (5 x 25 mg/d). Subsequently, AFI declined to 32. Upon cessation of indomethacin at 33 weeks, hydramnios resumed, followed by premature contractions. Amniotic fluid at delivery by cesarean section amounted to 5 l. The baby girl born (1690 g, Apgar 5/7/9) subsequently developed polyuria, hypokalemia and hypercalciuria. Otoacoustic emissions were normal. Urinary prostaglandin (PG) E2 excretion was largely elevated (371 ng/h/1.73m^2 [normal 4-27]), consistent with a diagnosis of a NKCC2 or ROMK kation transporter defect of the thick ascending loop of Henle (furosemide-type Bartter syndrome).

Amniotic fluid data:

	16 w	30 w	34 w
Indomethacin	off	on	off
Cl^- (mmol/l)	110	105	-
PG E2 (pg/mL)	-	19.8	130.1

Salt-losing nephropathies (hyperprostaglandin E syndrome) are rare causes of severe hydramnios. After exclusion of other underlying disorders, prolongation of pregnancy can be achieved by restricted amnioreduction and administration of high-dose indomethacin.