Lower urinary tract obstruction (LUTO) – prenatal intervention and long term outcome

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Abstract

Lower urinary tract obstruction in foetuses carries an untreated high rate of mortality. Ultrasound antenatal examination allows to introduce treatment in utero. Authors describe different therapeutics options and emphasize necessity of proper qualification to prenatal procedures. The role of antenatal therapy in LUTO was evaluated.

Key words: lower urinary tract obstruction, vesicoamniotic shunt, foetal cystoscopy

Introduction

Next to defects of the central nervous system and cardiac defects, urinary tract defects are the most commonly diagnosed pathologies during the prenatal period. Hydronephrosis – widening of pyelocalyceal system constitutes 50% of abnormalities in an abdomen observed during ultrasound examination of the foetus. The number of hydronephrosis diagnosed in utero is estimated for 1-3% of examined pregnant women. Sairam et al. examined 11465 women between 18 and 23 week of pregnancy and reported widening of pyelocalyceal system in 268 foetuses (2.3%). On the other hand, data obtained from Eurocat Working Group determined that the number of inborn hydronephrosis amounts to 1.5 cases among 10000 births [1, 2]. The most common cause of postnatally confirmed hydronephrosis is the narrowing (obstruction) within pelviureteric connection that constitutes up to 50% of defects, which in majority of cases do not require surgical treatment. Widening of urinary tracts caused by a bladder outlet obstruction constitutes about 10% of all diagnoses [3, 4].

The natural history of obstructive uropathy is variable and depends on numerous factors. In very severe cases obstruction of the urethra can lead to massive bladder distention, hydroureteronephrosis and renal fibrocystic dysplasia. Improper urine secretion to the amniotic cavity leads to oligohydramnios, and as a consequence causes pulmonary hypoplasia, deformation of extremities and face of the foetus. Condition of the baby after birth is determined by the degree of lung development and renal function. According to various sources death rate among children born with obstructive defect of the lower urinary tract may even amount to 54% [5, 6].

LUTO concerns innate defects causing obstruction of the male urogenital canal. Most commonly we are dealing with posterior urethral valve, prune-belly syndrome and urethral atresia. Other anomalies, including anterior urethral valve, metal stenosis or canal hypoplasia quite rarely constitute a problem diagnosed in the foetus. Bladder outlet obstruction in female gender may be caused by genitourinary sinus or cloacal abnormalities, and they often form a part of defect syndromes and stand as a completely different problem [7].

Prenatal diagnostics

LUTO is diagnosed on the basis of thorough ultrasound examination. All sections of the urinary tract have to be examined: condition of kidneys, their echogenicity, size of pyelocalyceal system, presence of cyst in the cortex, ureter width, size of the bladder and possible widening of urethra. This examination may initially suggest the etiology, level and severity of obstruction. Large, symmetric, round and thin walled bladder may reflect urethral atresia or urethral valves entirely blocking the lumen, urinary bladder of more tabular shape may indicate incomplete obstruction of urethra. The “keyhole sign” stands as a typical image reflecting bladder outlet obstruction.

Karyotype evaluation is also an important element. Due to oligo or anhydramnios amniocentesis may be impossible and the examination is performed through cordocentesis or chorionic villus sampling.

Foetal urine evaluation is another, extremely significant examination. Vesicoentesis and following urine analyses should be performed at least three times within 48h intervals (24-72 hours). Urine evaluation allows qualifying foetuses to a group with good or bad prognosis. (Table 1) [7-10].
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Intrauterine therapy

Bladder outlet obstruction hampering the proper urine passage through urethra causes secondary injury of all layers within the urinary tract. Widening the urethra above the obstruction, hypertrophy, hyperplasia and fibrosis of the bladder wall, thinning or hypertrophy of bladder wall with secondary urodynamic disorders, widening and prolonging the ureter and widening of the pyelocalyceal system as a result of improper urine flow to bladder or massive vesicoureteric reflux. All in all, renal dysplasia causes their improper functioning. Oligo or anhydramnios constitute the consequence of these changes, and this leads to improper pulmonary development and hypoplasia. Action aiming to enable proper urine outflow, bypass or cancel the obstruction as early as possible. Despite the fact that such procedures are widely acceptable, they constitute one of several indications towards intrauterine intervention within the scope of urinary tract, there still are many controversies related with qualifications, time and manner of prenatal actions. There are also critical reports on the influence of the shunting on the urinary bladder. Laboratory tests performed on sheep foetuses revealed that cystostomy implemented even to a healthy bladder causes reduction of its volume, lowering wall susceptibility, which may influence the postnatal functioning of the bladder [11]. Criteria qualifying the foetus for intrauterine therapy include: proper male karyotype, double-sided hydronephrosis, proper renal function (proper results of biochemical urine tests), and oligohydramnios. First experiences related with open foetal surgery were quickly abandoned. It seemed extremely promising that implanting shunt has to be strictly monitored. Initial controls should be performed every 24-48 hours, and then they can be limited to one examination a week or every two weeks, depending on the amount of amniotic fluid and condition of the foetus. Among possible complications we can enumerate shifting the drain outside the bladder, direct damage of the foetus (iatrogenic gastroschisis), bleeding from placenta and premature labour. Birth is performed according to standard obstetrical indications [7, 12-16].

Minimally invasive endoscopic techniques became a golden standard of procedure in many branches of medicine. Following the development of endoscopic techniques and device minimization, the use of endoscope also became possible in diagnostics and therapy of the foetus. The first attempt related with visualization of the human foetus in endoscopy has been undertaken as early as in 1954 [17]. Despite its name “minimally invasive” fetoscopy is a procedure of high invasiveness and requires significant experience and properly selected device. Current embryo-fetoscopes come with diameter 1.0-3.8 mm [18].

Such devices enable performing foetal cystoscopy through suprapubic puncture, accessing to posterior canal, determining the type of obstacle and in case of determining posterior urethral valve its mechanical ablation or ablation with the use of laser [19-21].

Treatment results

There are still a number of controversies concerning the efficiency of prenatal treatment with LUTO. What is taken into consideration when evaluating the results is mostly the survival after birth, the degree of pulmonary development and renal function. In 1997 Coplen presented results collected after analysing 169 foetuses with implanted shunt. 47% of children survived the birth and 40% of them developed renal insufficiency. He evaluated those complications after intrauterine intervention amounted to 45%. Nonetheless, within this group there was no division between foetuses with good and bad prognoses [22]. It has been determined that biochemical foetal urine parameters and lack of amniotic fluid during the initial period of pregnancy may cause death rate reaching even 95%. With proper selection and qualification for intrauterine therapy, results may be better and survival longer than 1 year may exceed even 90%. Shunting supporters emphasize that particularly in case of children with posterior urethral valve it shall be possible to improve renal function, delay the need of dialysis and transplantation. On the other hand other authors do not see any benefits in prenatal actions with postnatal primary ablation of the valve. Despite prenatal treatment of 50% of children low creatinine value during the first

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year of life stands as a very important factor prognosing renal function. All authors emphasize the role of precise qualification and the use of foetal cytostomy as well as intrauterine destroy of the valve to obtain proper results of foetal therapy. Increasing percentage of survival rate results in long-lasting treatment of large group of children. Quite often patients, who underwent prenatal diversion, require maintenance of ureterostomy or vesicostomy in order to improve the urine flow. In case of these children bladder augmentation procedures or creation of Mitrofanff fistulas are being performed, or it will be necessary to implement CIC in order to ensure proper bladder emptying. Only slightly more than 60% of shunted children have proper micturition. These disorders within lower urinary tract prove that only foetuses with greater injury of the urinary tract are being qualified for intrauterine procedures. Very severe problems are observed in children, who were shunted despite bad prognoses. The procedure was performed under pressure imposed by parents, patients survived the early postnatal period and they are potential candidates to dialysis therapy and renal transplant. Disorders within respiratory tract constitute a completely different problem. More than 40% of children suffer from asthma or recurring infections within urinary tract. What is more, 50% of children treated during the prenatal period are characterized by weight and height not exceeding 25 percentile. These complications are greatly related with patients suffering from prune belly syndrome, but they also occur in case of posterior urethral valve [13, 15, 18, 20, 23, 24].

Despite so many discrepancies relating the evaluation concerning prenatal treatment of patients with LUTO such proceedings seem extremely promising and they may result in significant improvement in therapeutic results among patient with this extremely severe complex of defects.

References


