NEPHROLOGICAL APROACH OF 5 CASES WITH NEURAL TUBE DEFECTS

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Abstract

Introduction: Neural tube defects result from the failed closure of the neural tube between the 3rd and 4th week of in utero development. Myelomeningocele represents the most severe form of dysraphism involving the vertebral column. *Objective:* The evaluation of prognosis and renal-urinary complications in children with myelomeningocele. *Material and method:* Authors present 5 cases with myelomeningocele and neuropathic bladder admitted in "Louis Turcanu" Pediatric Emergency Hospital Timisoara. We evaluated clinical aspects, pathogenesis, evolution, complications and the treatment of these cases. *Results:* In all cases the defect was surgically corrected. In two cases shunting procedure for hydrocephalus followed, while the other cases present spontaneous stabilization of hydrocephalus. Mental retardation was sever (1 patient), moderate (1 patient) and mild (3 patients). Urinary anomalies consisted of neuropathic bladder (5 cases), massive bilateral hydronephrosis (4 patients) and horseshoe kidney (1 patient). The patients presented urinary incontinence, urinary infections (1 case deceased from sepsis) and renal failure. The immobilization, urinary and fecal incontinence represent a sever handicap for these patients and their family. Social integration is difficult. Patients' prognosis is unfavorable: 1 case deceased at six years of age, two cases already with renal failure and the recurrence of urinary tract infections followed, probably, by renal failure for the other two patients. *Conclusions:* The association of genitourinary system pathology determined an unfavorable evolution and a negative prognosis for these cases. Folic acid supplementation should be initiated before conception and continued until at least 12th wk of gestation in order to prevent the neural tube defects.

Key words: myelomeningocele, neuropathic bladder, prognostic

Introduction

Neural tube defects result from the failed closure of the neural tube between the 3rd and 4th week of in utero development. Myelomeningocele represents the most severe form of dysraphism involving the vertebral column.

The pattern of inheritance of these malformations is multifactorial, rendering the identification of the underlying causes. Essential signaling pathways of the development of the central nervous system include the planar cell polarity pathway, which is important for the initiation of neural tube closure as well as well as sonic hedhehog pathway, which regulates the neural plate bending.[1]

Recent findings suggested a link between cilia and the planar cell polarity signaling cascade. In particular, on focus on how this interaction may influence the process of neural tube closure and how these results may be relevant to our understanding of common human birth defects in which neural tube closure is compromised.[2]

Genetic studies in NTDs have focused mainly on folate-related genes based on the finding that perinatal folic acid supplementation reduces the risk of NTDs by 60-70%.[3]

Risk factors for renal injury in patients with meningomyelocele are: increasing age, evidence of hydroureteronephrosis and vesicoureteric reflux, high leak pressures and low bladder volume.[4]

Objective

The authors evaluated prognosis and renal-urinary complications in children with myelomeningocele.

Material and methods

Authors present 5 cases admitted in "Louis Turcanu" Pediatric Emergency Hospital Timisoara with diagnosis:

- ❑ Neural tube defects
- Myelomeningocele
- Stabilizated hydrocephalus
- Cerebral palsy
- Neuropathic bladder
- Urinary incontinence
- ❑ Reno-urinary anatomic or functional anomalies

We evaluated clinical aspects, pathogenesis, evolution, complications and the treatment of these cases.

Results

I Patriciu, 2 years

- ❑ Neural tube defects
- Myelomeningocele
- Stabilizated hydrocephalus – ventriculoperitoneal shunt
- Arnold Chiari II malformation
- Cerebral palsy
- Neuropathic bladder
- Urinary and fecal incontinence
  - ❑ Ureterohydronephrosis
  - ❑ Right vesico-ureteral reflux - ureterostomy
  - ❑ Recurrent urinary tract infections
  - ❑ Growth retardation

Table 1 Recurrent urinary tract infections of I Patriciu

Days	198	275	379	645
Urine culture	E coli	Klebsiella	E coli	Contamination

N Iulia, f, deceased at 6 years of age from sepsis with MSOF

- ❑ Neural tube defects
- Myelomeningocele
- Stabilizated hydrocephalus – ventriculoperitoneal shunt
- Cerebral palsy
- Neuropathic bladder
- Urinary and fecal incontinence
  - ❑ Horseshoe kidney with secondary hydronephrosis
  - ❑ Vesico-ureteral reflux
  - ❑ Recurrent urinary tract infections
  - ❑ Growth retardation

Table 2 Recurrent urinary tract infections of N Iulia

Days	1001	1082	1105	1198	1342	1507
Urine culture	Proteus	Pseudomonas aeruginosa	Pseudomonas aeruginosa	Pseudomonas aeruginosa + E coli	Pseudomonas aeruginosa	Negative

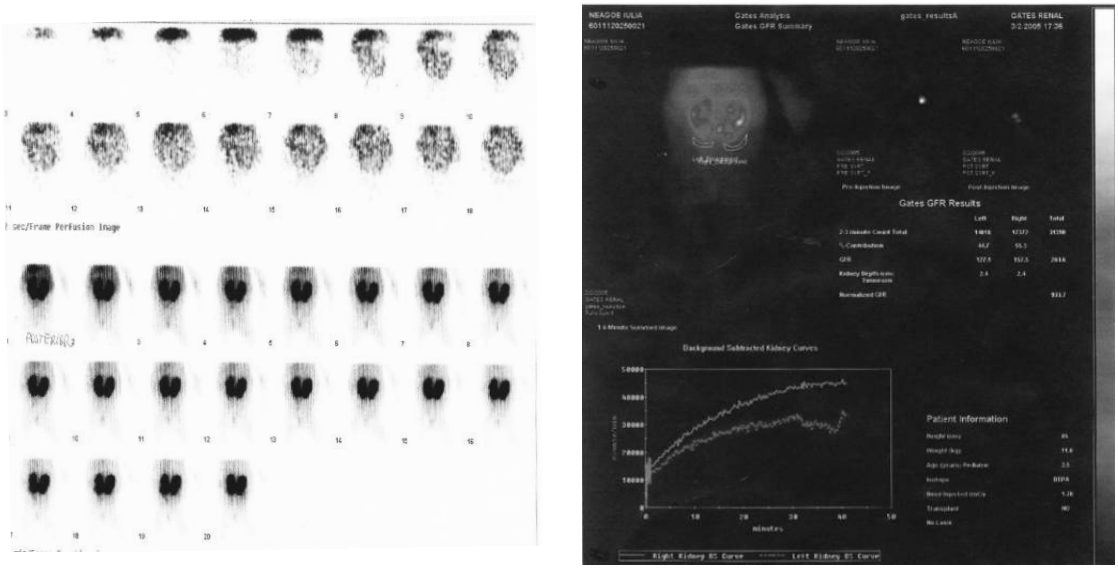


Figure 1a, b N Iulia's Tc-DMSA planar renal scintigraphy show bilateral impairment of renal function.

I Dragana, f, 14 years

- ❑ Neural tube defects
- Myelomeningocele
- Stabilizated hydrocephalus
- Cerebral palsy
- Neuropathic bladder
- Urinary and fecal incontinence
  - ❑ Ureterohydronephrosis
  - ❑ Recurrent urinary tract infections

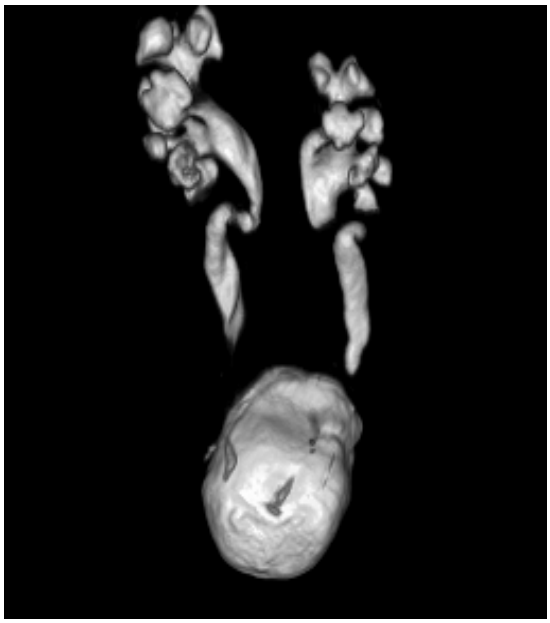
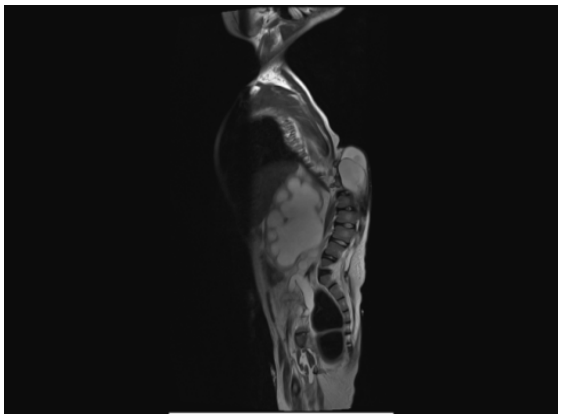


Figure 2 I Dragana's MRI: neuropathic bladder and bilateral ureterohydronephrosis.

S Ionut, m, 15 years
- ❑ Neural tube defects
- Myelomeningocele
- Stabilizated hydrocephalus
- Cerebral palsy
- Neuropathic bladder
- Urinary and fecal incontinence
  - ❑ Ureterohydronephrosis
  - ❑ Recurrent urinary tract infections
  - ❑ Chronic renal failure - diagnosed at 15 years of age

No compliance treatment and follow up.



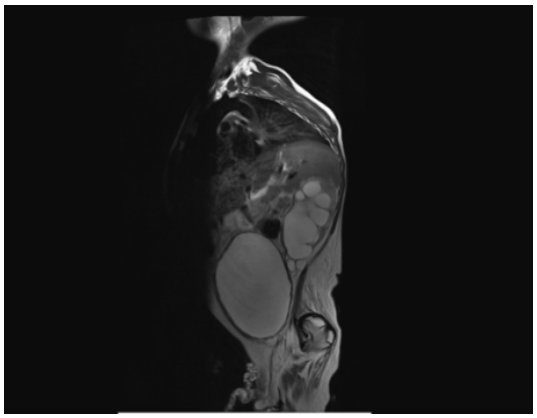


Figure 3a, b S Ionut's MRI: neuropathic bladde and bilateral ureterohydronephrosis.

C Adina, f, 14 years
- ❑ Neural tube defects
- Myelomeningocele
- Stabilizated hydrocephalus – ventriculoperitoneal shunt
- Cerebral palsy
- Neuropathic bladder
- Urinary and fecal incontinence
  - ❑ Ureterohydronephrosis
  - ❑ Recurrent urinary tract infections
  - ❑ Chronic renal failure - discovered at 4 years of age
  - ❑ Renal anemia – treated with Erythropoetin

No compliance to treatment and follow up – 10 years without medical follow up.

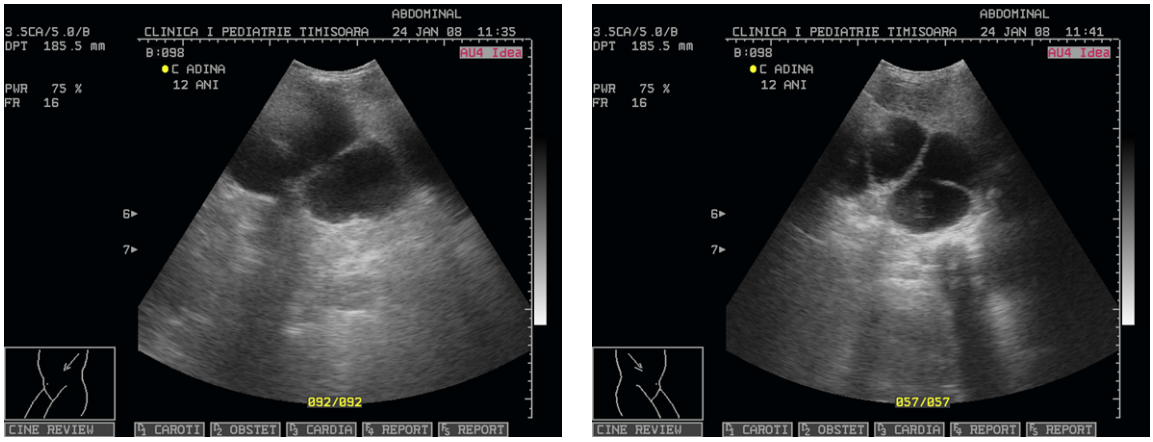


Figure 4a, b C Adina's renal ultrasound show bilateral ureterohydronephrosis.

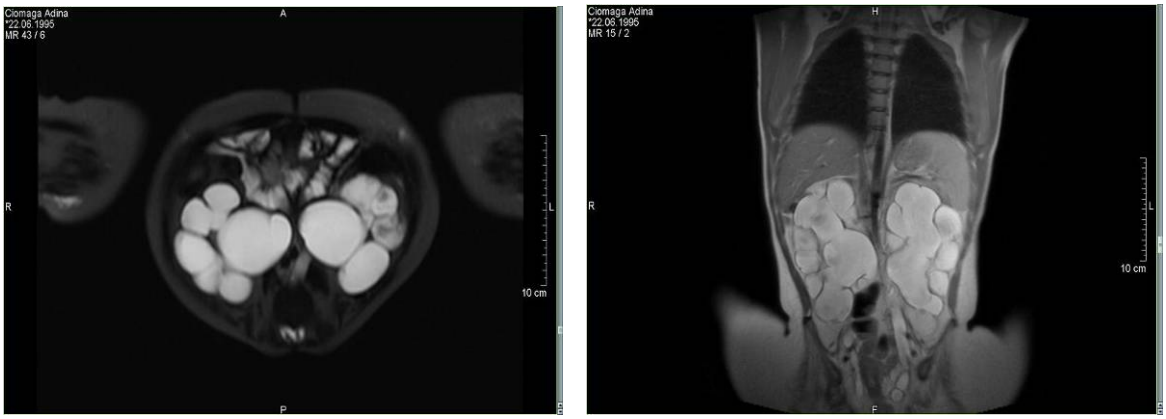


Figure 5a, b C Adina's MRI: neuropathic bladder and bilateral ureterohydronephrosis.

Discussions

In all cases the defect was surgically corrected. In two cases shunting procedure for hydrocephalus followed, while the other cases presented spontaneous stabilization of hydrocephalus. Mental retardation was sever (1 patient), moderate (1 patient) and mild (3 patients).

Uroneurological assessment of these patients must be performed repetitive.[5] Urinary anomalies consisted of neuropathic bladder (5 cases), massive bilateral hydronephrosis (4 patients) and horseshoe kidney (1 patient).

The patients presented urinary incontinence, urinary infections (1 case deceased from sepsis) and renal failure (2 patients).

The common goal in caring for these patients must be the prevention of progressive renal damage. However, once kidney failure has occurred, good and safe techniques for renal replacement therapy[6] are available to bridge the time to transplantation, which is undoubtedly the best treatment for these patients.[7]

The immobilization, urinary and fecal incontinence represent a sever handicap for these patients and their family. Social integration is difficult. The pubertal development of this patient may lead to improving there urodynamics.[8]

Patients' prognosis is unfavorable: 1 case deceased at six years of age, two cases already with renal failure and the recurrence of urinary tract infections followed, probably, by renal failure for the other two patients.

Conclusions

The association of genitourinary system pathology determined an unfavorable evolution and a negative prognosis for these cases.

Folic acid supplementation should be initiated before conception and continued until at least 12th wk of gestation in order to prevent the neural tube defects.[9,10]

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