Uro-nephrological Outcome of Patients with Neural Tube Defects: Would a Spina Bifida Clinic Make a Difference?

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Abstract

Aim of work: Review our experience over the last 10 years for our patients with myelomeningocele (MMC), followed in our Spina Bifida Clinic, in particularly looking at the uro-nephrology outcome for those patients.

Material and Methods: We retrospectively reviewed the medical records of all patients followed at the combined Spina Bifida Clinic between 1999 and 2009, and had at least one year of follow-up with us.

We looked at their demographic data, uronephrological status at presentation and at the latest follow up, and the rate of surgical intervention.

Results: 188 patients were in active follow up during the 10 years period. Mean age at presentation was 5.3 years +/- 3.6 SD. One hundred and nine (58%) were on clean intermittent catheterization, 44 had Botox injection and 26 had bladder reconstruction at their last follow up. Sixty six percent of the patients presented to us older than 3 year, where this group had a significantly higher rate of surgical intervention Botox or reconstruction) compared to those who came early to us (P = 0.003 & 0.025)

Conclusion: Multidisplinary Spina Bifida Clinic is an integral part in management of patients with MMC, in order to reach a safe urological outcome.

Early presentation to our clinic resulted in lesser need for surgical intercession compared to those presented later than 3 years of age.

Key words: Spina Bifida, urological outcome, Saudi Arabia, Botullinum toxin

الملخص:

الهدف من العمل: إستعراض تجربتنا خلال السنوات العشر الماضية لمرضانا الذين يعانون من الفتق السحائي ، والذين تتم متابعتهم في عيادة خاصة بذلك ، وبخاصة نتائج المتابعة الخاصة بأمراض الكلى والمسالك البولية لهؤلاء المرضى.

طريقة البحث: إستعرضنا بأثر رجعي السجلات الطبية لجميع المرضى في عيادة الفتق السحائي المشتركة بين عامي 1999 و 2009 ، والذين لاتقل فترة متابعتهم معنا عن سنة واحدة ، وقمنا بالبحث في بياناتهم الديموغرافية ، وحالة الكلى والمسالك في أول زيارة ، وعند آخر متابعة ، وحاجة هؤلاء المرضى للتدخل الجراحي .

النتائج: يوجد 188 مريضاً في متابعة مستمرة خلال فترة 10 سنوات ، وكان متوسط العمر 5,3 سنة " ± 3,6 " . وفي آخر متابعة كان هناك مائة وتسعة مرضى (58%) يستخدمون القساطر المتقطعة لإخراج البول ، و44 مريضاً قد تم حقنهم بالبوتوكس و 26 مريضاً إحتاجوا إلى إعادة تهيئة للمثانة البولية . كان 66% من المرضى اكبر من 3 سنوات عند الفحص الأول وقد بلغت النسبة الأكبر فيهم في حاجة إلى التدخل الجراحي بإستخدام البوتوكس أو إعادة تهيئة المثانة البولية بالمقارنة بأولئك الذين جاءوا الينا في وقت مبكر .

الخلاصة: عيادة الفتق السحائي المشتركة هي جزء لايتجزأ في متابعة المرضى الذين يعانون من هذا المرض ، من أجل التوصل إلى نتائج آمنه للمسالك البولية ، تؤدي المتابعة في وقت مبكر في عيادة الفتق السحائي المشتركة غلى حاجة اقل للتدخل الجراحي مقارنة مع تلك التي قدمت في وقت متأخر بعد 3 سنوات من العمر .

INTRODUCTION:

Myelomeningocele (MMC) affects 1 in 1000 births, comprising the most common cause of pediatric neuropathic bladder. This group of patients usually suffering of varying degree of disabilities necessitating the need for a multidisciplinary team to take care of those patients including pediatric urologist, neurosurgeons, orthopedists as well as specialized nurses and social workers.[1]

Urological manifestation of patients with MMC is quite common, with a signified psychological and medical negative effect on those patients, which mandate early follow-up and a comprehensive management to prevent any irreversible renal damage and stabilizes the bladder function.[2]

Accordingly, the urological care of those patient is one of the essentials of establishing a Spina Bifida Clinic, as urological care of children with spina bifida has undergone several important changes in the last two decades.[3] We established the Spina Bifida Clinic at King Khalid University Hospital in 1999, currently it is a bimonthly clinic, with 188 patients are actively following.

Here we review our experience over the last 10 years for our patients with MMC followed in the Spina Bifida Clinic and emphasize on the uro-nephrology outcome for those patients.

MATERIALS AND METHODS:

We reviewed the files of all patients followed at our multidisciplinary Spina Bifida Clinic between 1999 and 2009 and had at least one year of follow-up. All our patients are followed at our bimonthly Spina Bifida Clinic, where referrals are either accepted from children delivered at our institution or from other centers in the Kingdom.

The patients were evaluated by the urology team where a baseline renal ultrasound, urine test and renal profile requested. All patients by the age of 6 months (or at presentation if referred to us after the age of 6 months) will have a urodynamic study to evaluated their bladder. If the kidneys are dilated (hydronephrotic), or urodynamic studies reveal a non compliant bladder or urinary tract infection was documented, a voiding cystogram (VCUG) is performed.

Patient age at presentation, period of follow-up, demographic data, uronephrological status at presentation and at last follow up, current urological active management including surgical interventions, upper tract status and renal profile early and at last follow up were evaluated in the current study.

Upper tract deterioration was defined as radiographic evidence of ureteral dilatation or hydronephrosis on ultrasound, or new onset or progressive vesicoureteral reflux on VCUG. Abnormal bladder compliance was defined as intravesical pressure during filling or at time of leakage exceeds 40 cm H20.[4]

Active management will start according to the above mentioned evaluation and baseline investigation, (Table 1).

RESULTS:

Currently, we have 188 patients who had active follow-up in our Spina Bifida Clinic and had at least one year of follow-up in King Khalid University Hospital. The mean age at presentation to our clinic was 5.3 years +/- 3.6 SD (range between 0.2 to 14 years), there were 97 male (51.5%) and 91 female (48.5%). Most of the families are living in Riyadh (125 [66.5%]). Mean follow up period was 3.6 years +/- 1.9 SD (1-10 years).

Out of 188 patients, 109 (58%) are on CIC + anticholinergic medications either due to non compliant bladder and/or urinary incontinence. However, 79 patients (42 %) show stable bladder function, upper tract and renal functions and they are under close follow up observations.

Out of the 109 patients, 44 (40.3%) patients had refractory urinary incontinence and/or their upper tract didn't improve with the conservative management, accordingly, they were moved to be managed with intradetrusor Botox injections.

Out of the 109 patients, 26 (23.8%) patients had bladder reconstruction at their last follow-up, 16 had Botox before and 10 had surgical intervention before we started this Botox program (Table 2).

At presentation 160 (85.1%) patients had normal renal function with mean serum creatinine of 46.6 +/- 40 SD and in the last follow up 178 (94.6%) have normal renal function with mean serum creatinine of 41.6 +/- 32 SD.

Out of 188 patients 63 (33.5%) patients presented to us before the age of 3 years and 125 (66.5%) presented older than 3 years.

The older age group was significantly higher in the rate of surgical interventions than the younger group. From the older age group, 37 patients underwent Botox injection and 22 were subjected to reconstructive surgery in comparison to 6 and one patient respectively for those who presented before the age of 3 years (P value is 0.003 and 0.025 respectively)

DISCUSSION:

The goals of urologic care and management of children with neural tube defects is to maintain a normal renal functions and gaining normal social life by maintaining their urinary and stool continence.[5] This is the aim of the Urology group as part of the Spina Bifida Team. Accordingly, all patients once seen by the urology team a comprehensive management protocol is stated, urotherapy nurse introduces the management protocol to the family, and base line investigations is done, management will then follow according to the schedule in Figure 1.

Fifty eight percent of our patients were in need of clean intermittent catheterization either to manage the high intravesical pressure or to gain social continence; we had an excellent outcome from our CIC Program[6] despite the social difficulties that we face initially. Medical management with CIC and anticholinergics is effective in preserving renal function and providing safe urinary continence in however, this therapy fails in approximately 10-15% of patients for whom surgical reconstruction is the only valid option.[7] Recently, intradetrusor injection of Botulinum toxin- A (Botox) proved its efficacy as 2nd line of treatment for those group of patients.[7,8]

In our group of patients, medical management with CIC (Fig 1)and anticholinergics was sufficient in preserving renal function and providing safe urinary continence in 55 patients (out of the 109) and surgical interventions were indicated in 54 out of the 109 (28% of the total group of patients. This might be explained by the late presentation of our patients as about 66.5% of our patients presented to our clinic at age of 3 years or older.

Before 2003, surgical reconstruction was the solution for those patients not responding to medical managements; however, in 2003 we started our experience with Botox injection for that group of patients not responding or have side effects to medical treatments. Out of the 188 patients, 44 (23.4%) patients underwent Botox injection, and 23 of them had a good response after repeated injections and we were apple to avoid major surgical reconstruction for them.

Rate of surgical reconstruction (Figure 2) in our patients was 26(13.8%), 10 patients before implementation of Botox program and 16 patients after unsuccessful trial with Botox injection.

Out of 26 patients underwent surgical reconstruction, 22(86%) patients presented to us at age of 3 years or older which is significantly higher (P=0.025) than patients presented at younger age (0 to 3 years) where only 4 patients from this group were in need for surgical reconstruction.

At the same time, out of 44 patients for whom Botox injection were indicated, 37(84%) were from the older age group which is also significantly higher (P=0.003) than the younger age group where only 7(16%) patients underwent Botox injection.

This rate of surgical interventions needed were much lower with early presentation and early start of treatment and this support the need for early proactive management[9] for those patients accompanied with strict, regular and well constructed program to avoid the high rate of surgical interventions and associated complications.

CONCLUSION:

A safe kidney function with social continence can be gained in most if not all patients with NB secondary to MMC if they are properly followed with a dedicated team in a Spina Bifida Clinics, although the follow-up in quite exhausting. We recommend early follow for those patients, together with early start of CIC program if needed.

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Figures:

Figure1: Pictures describe teaching sessions of CIC on models in our urotherapy laboratory.

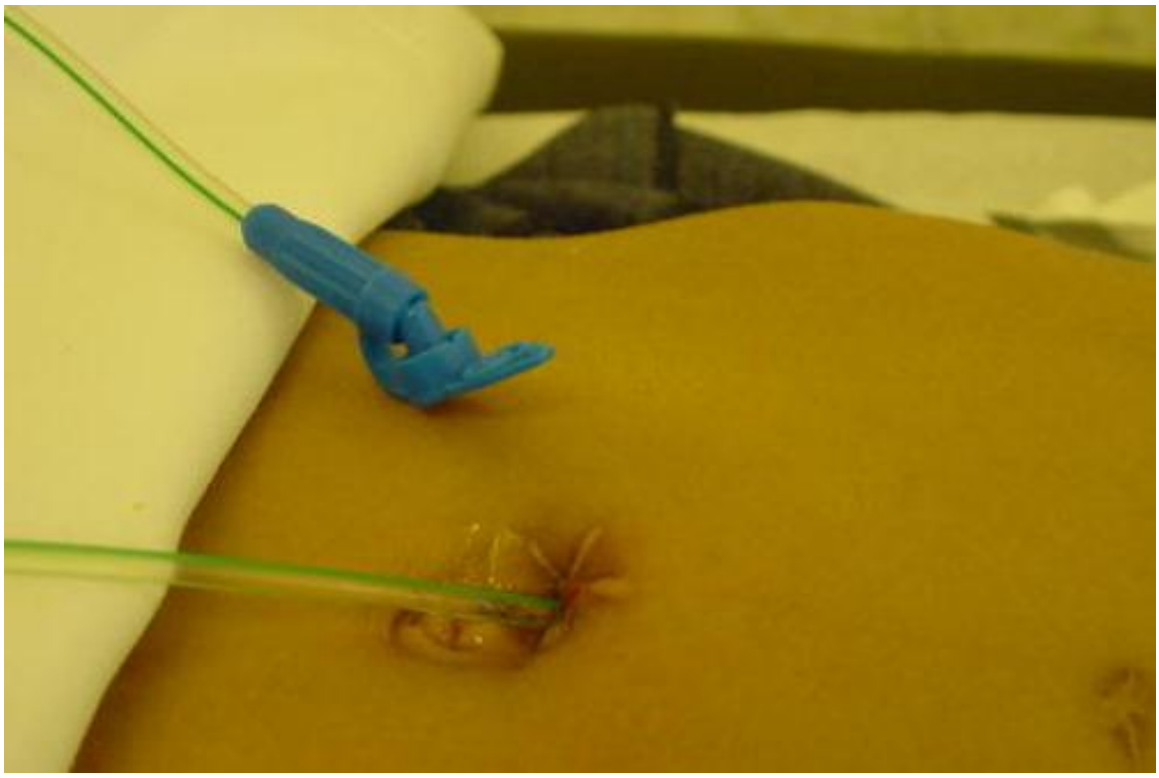

Figure 2: patient post ileocystoplasty and mitrofanoff procedure doing CIC for himself