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Investigation into neurogenic bladder in arthrogryposis multiplex congenita

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Abstract *Objective:* During the follow-up of children who had been diagnosed with arthrogryposis multiplex congenita (AMC), it was noted that some were experiencing dysfunctional voiding. Further investigation into these cases led to a diagnosis of neurogenic bladder. Few studies have investigated the relationship between AMC and neurogenic bladder, this being the first to describe the clinical characteristics of neurogenic bladder among these patients.
Methods: A series of 26 cases were obtained from the electronic medical records of patients with AMC who were admitted to Hospital Sarah in Salvador between 1994 and 2007. The patients had all been diagnosed with neurogenic bladder through clinical symptoms, lower urinary tract exams, and urodynamic findings.
Results: There was urinary incontinence in 21 patients (81%), and 50% had a history of urinary tract infections. Renal function was altered in 4 patients (15%) and normal in 22 (85%). In the urodynamic study, 14 patients (64%) had detrusor overactivity and 6 (27%) had underactivity.
Conclusion: Patients with AMC may show changes in the urinary tract, including neurogenic bladder. It is mandatory to study these symptomatic children with urinary disorders.

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Introduction

Arthrogryposis multiplex congenita (AMC) is defined as a non-progressive congenital rigidity disorder that affects multiple joints [1,2]. There are around 150 symptoms that are associated with multiple congenital contractures. Congenital amyoplasia, the most common type of congenital arthrogryposis, usually results from impaired blood flow to the placenta and leads to the death of the embryo's marrow [3—5].

There are a few publications that establish a correlation between arthrogryposis and genitourinary changes, but not with respect to a possible association with neurogenic bladder [6]. There is a description of concomitance between arthrogryposis and the malformation of the genitourinary system, as in cryptorchidism, hypospadias, renal agenesis and lithiasis. The presence of neurogenic bladder among patients with arthrogryposis has been reported, but without a description of its characteristics [7,8].

The objective of this study was to describe the urinary symptoms and the results of urological examination of patients with neurogenic bladder and AMC.

Materials and methods

A series of cases were analyzed in order to describe the characteristics of neurogenic bladder in patients with arthrogryposis. Between March of 1994 and January of 2007, a total of 226 patients diagnosed with AMC were admitted to Hospital Sarah of Salvador, Bahia, Brazil.

All patients with AMC who showed lower urinary tract symptoms underwent urodynamic study to investigate for neurogenic bladder, independent of their age when admitted to the hospital. The criteria for exclusion were: patients without urological data on electronic medical records, or if urodynamic study was done in another institution, but without a proper description in the electronic medical records, and patients with other associated conditions of arthrogryposis such as spinal dysraphism, hydrocephalus, cerebral palsy, mental retardation, or paraparesis of other etiology. Fig. 1 summarizes the selection of patients.

The data were obtained by review of the electronic medical records of each patient, following a specific protocol.

The different levels of impairment were categorized with relation to the arthrogryposis classification according to Judith Hall [9]:

Type I — Predominantly limb-related disorders (amyoplasia, distal forms of arthrogryposis).

Type II — Disorders involving the limbs as well as some other body parts.

Type III — Disorders that involve the limbs as well as central nervous system (CNS) dysfunction.

The clinical manifestations investigated were: urgency, daytime incontinence, nocturnal enuresis, and the presence of a urinary tract infection as well as recurrent urinary tract infections. The exams included renal function (through both urea and creatinine tests), kidney and urinary tract ultrasounds, voiding cystourethrography, and urodynamic studies. During the urodynamic study (Medtronic Duet, Minneapolis, MN, USA), uroflowmetry was initially carried out, followed by cystometry and pressure flow studies. An anal electrode was used to record the electromyographic data of the external urethral sphincter and perineal muscles. Methods and definitions of urodynamic evaluations followed the guidelines of the International Continence Society [10]. The treatment methods were: guidance on voiding at scheduled times and intermittent bladder catheterization with or without the use of anti-cholinergic drugs.

Statistical analysis

The data were analyzed using the statistical program SPSS v16.0 (SPSS, Chicago Inc., IL, USA). The program analyzed the mean, standard deviation and frequency of each variable.

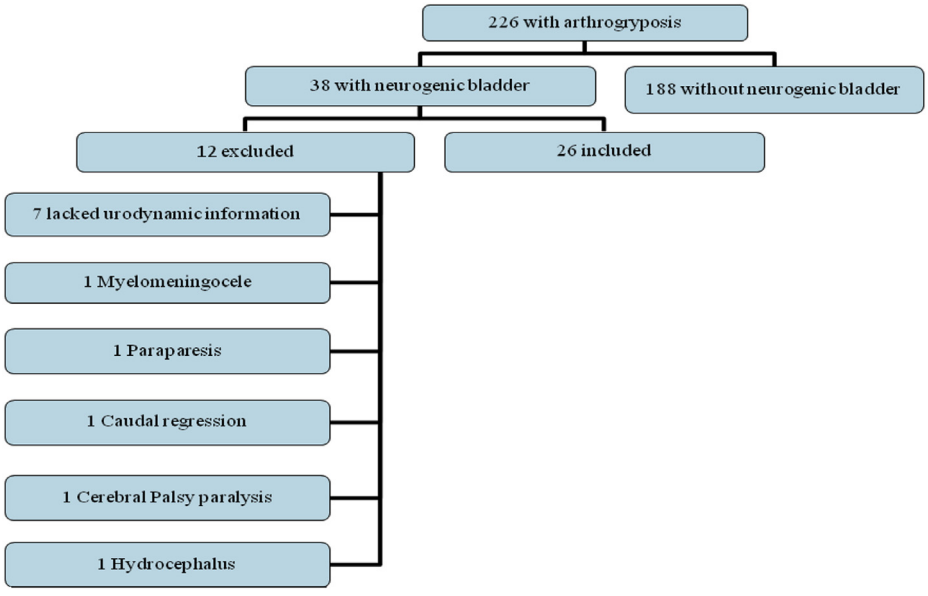

Figure 1 Patient selection process.