Reviews in Paediatric Urology

edited by J. HERBERT JOHNSTON

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1974

EXCERPTA MEDICA, AMSTERDAM

Library of Congress Catalogue Card Number 73-88154
ISBN Excerpta Medica: 90 219 2085 9
ISBN American Elsevier: 0444 15 153 2

PUBLISHERS
Excerpta Medica, Amsterdam

SOLE DISTRIBUTORS FOR THE U.S.A. AND CANADA
American Elsevier Publishing Company, Inc.
52 Vanderbilt Avenue, New York, N.Y. 10017

Printed in The Netherlands by Drukkerij Mouton Co., The Hague

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Introduction

That is the worst of learning from experience; it takes too long. Often it takes a lifetime. "Experience," said Sainte-Beuve, "is like the pole star; it only guides a man in the evening, and rises when he is going to rest."

Reginald L. Hine. In Confessions of an Uncommon Attorney.

Paediatric urology has but fairly recently emerged from being the interest of a very few surgeons working in a highly specialised environment to being a subject of great clinical importance to the practising urologist and also to acquiring the status, in an increasing number of academic centres throughout the world, of being a specialty in its own right. The last few years have seen new developments and great advances in our knowledge of childhood disorders of the genito-urinary tract, both congenital and acquired. There are, however, still many areas where our comprehension is severely limited and where, as a consequence, disagreements, sometimes profound, exist especially as regards optimal management.

In this book, which it is hoped may be the first of a series, we have selected some important aspects of paediatric urology for review and our aim is to provide the post-graduate student with a distillate of the extensive experience of authors who have had a particular interest in their various subjects.

We thank the contributors for their enthusiastic acceptance of their tasks and for their diligence in the preparation of their chapters.

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Contents

Distal urethral stenosis

R. P. LYON

The distal urethral ring, composed of collagen, is the narrowest segment of the lower urinary tract in the female child. Because of its relatively small diameter and its inflexibility, the ring has been termed *distal urethral stenosis*. It causes enough urethral discomfort to trigger sphincteric muscle spasm, setting the stage for inhibited and incomplete voiding, the latter providing residual urine susceptible to infection. With dilation of the ring to larger and sustained diameters, the signs and symptoms of difficult urination are relieved.

Historical background

Radical changes in the therapy of girls with urinary tract infection have occurred since recognition of the distal urethral ring (Lyon and Smith, 1963). At that time, the bladder neck was the point of attack as the obstructive agent. Although some urologists claimed excellent results with plastic surgical procedures designed to enlarge the bladder neck, the majority felt much as did David Williams (1958b), who in discussing bladder neck obstruction wrote that "the clinical ... and the pathological diagnosis has been made by the process of exclusion". It was disquieting to the urologic surgeon as he carried out his Y-V plasty of the bladder neck not to find a thickened bladder musculature as testimony to active

obstruction and to find later that surgical cure had not been achieved.

In the late 1950's, I attempted to make a more determined effort to study the effects of a Y-V plasty of the bladder neck on girls with serious urinary tract infections. It seemed clear that assessment of urinary flow would be the most critical measure of the effectiveness of the therapy (Lyon and Smith, 1963). To be reliable and reproducible, any test would have to be as free as possible of devices that would psychologically inhibit normal flow. The ordinary tape recorder, with pickup attached to the office commode, proved to be the answer. More sophisticated methods have since been developed (Keitzer, 1966), such as the conversion of sounds to tracings as with an EKG, but our experience suggests that the ear is still the most acute judge of flow patterns and certainly offers the most available method of making such measurements.

In order to determine whether Y-V plasty at the bladder neck changed the flow pattern, I made pre- and postoperative recordings in three cases. In two of the three, voiding patterns did not change from the initial intermittent stream and residual urine remained high. Although the bladder neck could be an agent of obstruction, it lost its credentials as *the agent* in two of these cases.

Still hoping to give the bladder-neck theory another chance, we adapted the bougie á boule to urethral calibration in the hopes that as it was passed through the urethra a difference in muscular thickness could be palpated through the rectal wall and perhaps a thick bladder neck identified (Lyon and Smith, 1963). It was at this point that the distal urethral ring was recognized, for even bougies as small as F16 were not admitted through the distal urethra in many cases.

Anatomy

The inner longitudinal muscle fibers of the detrusor continue as the inner longitudinal fibers of the urethra, exerting the least closure effect (Tanagho and Smith, 1966). The middle circular fibers of the detrusor do not enter the urethra. The outer longitudinal fibers of the detrusor continue into the urethra as the outer circular or oblique coat of fibers and supply a greater urethral sphincteric action. Surrounding these fibers and probably interdigitating with them are the strands of the striated external sphincter, a part of the perineal musculature. These muscles exert the greatest closure effect throughout the distal two-thirds of the urethra and play a major part in the maintenance of continence.

Both the inner and outer layers of urethral musculature appear to

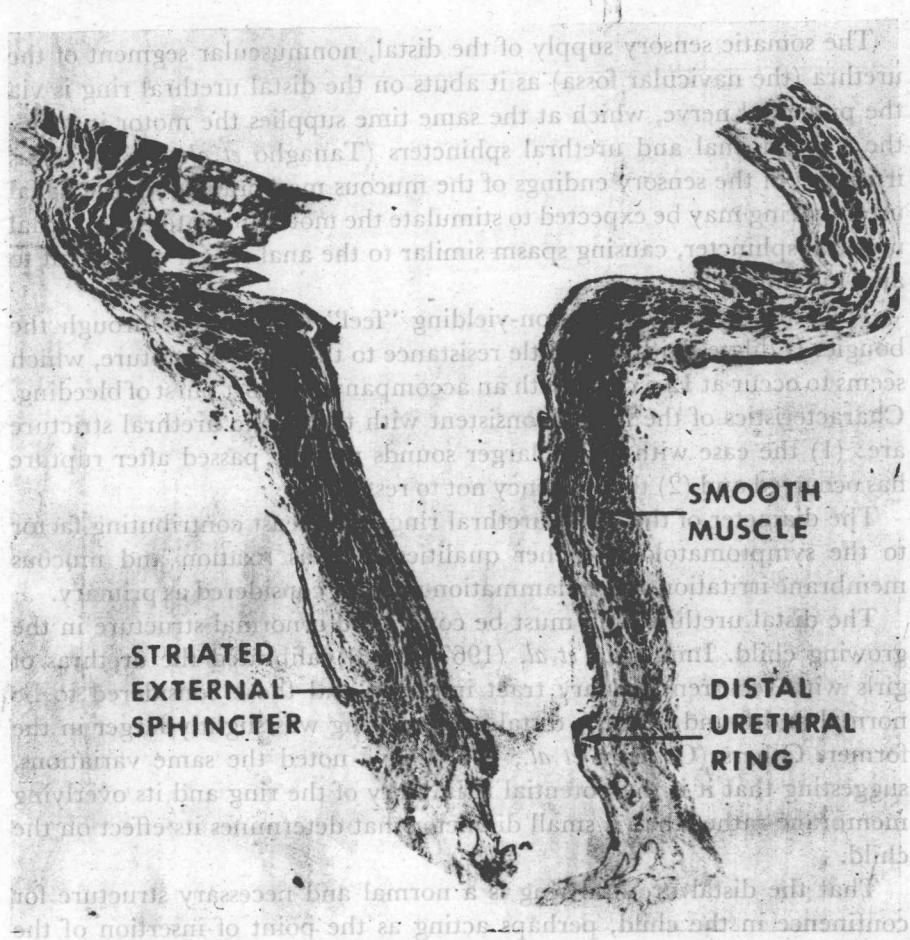

Fig. 1. Sagittal section of urethra with distal urethral ring. Modified from Lyon and Tanagho (1965).

insert into a dense, collagenous tissue ring situated in the distal urethra and termed the distal urethral ring (Fig. 1). The collagenous tissue continues down to the external meatus with some elastic fibers interdispersed. The condensation of collagenous tissue in the distal third of the urethra results in a narrow, minimally stretchable segment. This overabundance of collagen tends to encroach upon and indent the lumen of the distal urethral segment, a histological finding that is thought to be the anatomical basis of the ring. The depth of the ring seems to depend on the length of the nonmuscular segment. Thus the ring may be almost at the meatus or as deep as 1 cm proximal to it.

The somatic sensory supply of the distal, nonmuscular segment of the urethra (the navicular fossa) as it abuts on the distal urethral ring is via the pudendal nerve, which at the same time supplies the motor input to the striated anal and urethral sphincters (Tanagho *et al.*, 1971). Thus irritation of the sensory endings of the mucous membrane over the distal urethral ring may be expected to stimulate the motor fibers to the external urethral sphincter, causing spasm similar to the anal spasm coincident to anal inflammation.

The ring has a fibrous, non-yielding "feel" transmitted through the bougie. It dilates with very little resistance to the point of rupture, which seems to occur at 12 o'clock with an accompanying short burst of bleeding. Characteristics of the ring inconsistent with the classic urethral stricture are: (1) the ease with which larger sounds may be passed after rupture has occurred and (2) the tendency not to restenose.

The diameter of the distal urethral ring is the least contributing factor to the symptomatology. Other qualities such as fixation and mucous membrane irritation and inflammation must be considered as primary.

The distal urethral ring must be considered a normal structure in the growing child. Immergut *et al.* (1967; 1968) calibrated the urethras of girls with recurrent urinary tract infection and those considered to be normal and found that the distal urethral ring was slightly larger in the former. Others (Graham *et al.*, 1967) have noted the same variations, suggesting that it is the potential irritability of the ring and its overlying membrane rather than a small diameter that determines its effect on the child.

That the distal urethral ring is a normal and necessary structure for continence in the child, perhaps acting as the point of insertion of the urethral musculature, is demonstrated by its absence to bougie palpation in four youngsters under our care. Two would be considered classic female epispadiases, with bifid clitori; two have short urethras. All are incontinent to some degree.

The distal urethral ring appears to be influenced by maternal estrogens at birth and by the child's own hormonal changes at the menarche. Fischer *et al.* (1969) calibrated the urethras in newborns and found an average diameter of 16F, a larger diameter than that found at ages one and two (Immergut *et al.*, 1967). Williams (1958a) also noted that the female urethra at birth will admit a somewhat larger instrument than at the age of one or two, presumably a result of maternal estrogen influence on the ring.

The ring tends to regress at the menarche. Our office calibrations of teenagers seen with first infections, post-coital in nature, demonstrate a sharp increase in the distal urethral diameter at ages 14 to 18 out of

proportion to the overall growth curve. This observation is corroborated by the figures of Immergut in his "normal" series (Immergut and Wakeman, 1968). In the majority of teenagers, the exact position of the distal urethral ring when present is hard to judge, for its gritty quality is absent.

Pathological physiology

The diameter of the ring itself is rarely small enough to diminish urinary flow below a normal rate (20 cc/sec in the presence of a normal detrusor contraction (40 to 50 cm H_2O pressure) (Ritter *et al.*, 1964). Hydraulic laws governing conduit flow absolve a ring greater than F12 of limiting normal flow. The distal urethral ring rarely calibrates smaller than this. Thus diminished flow rates and interrupted voiding patterns must be explained otherwise. This variable resistance to flow, assuming a constant and smooth detrusor contraction, is clearly heard on the tape when compared to the smooth and normal-to-high flow rate patterns produced by children asymptomatic after a single intensive urethral dilation under anesthesia.

Although voiding urethrograms had suggested the mechanism of variable obstruction to be that of striated sphincter spasm (Lyon and Tanagho, 1965) more sophisticated proof was needed. It was provided by Tanagho *et al.* (1971), who applied a proven anatomical and physiological expertise to the problem. By means of intraurethral and intravesical pressure measurements, using a modified Enhörning set of tiny balloons, he studied girls with a symptomatic distal urethral ring, some without infection and others with recurrent lower urinary tract infection refractory to both medical treatment and urethral dilation. Normal resting urethral pressure reaches its highest sustained level throughout the distal two thirds of the urethra, proximal to the distal urethral ring, presumably delineating the area of striated sphincter muscle activity (Fig. 2). In children with a symptomatic distal ring, abnormally high resting pressures were recorded in the sphincteric segment. These pressures dropped to normal temporarily with the use of skeletal muscle blocking agents such as curare. Voiding was repeatedly interrupted by irregular bouts of activity and frequently was terminated with residual urine of 5 to 20 cc. Concomitant urethrography disclosed great variations in urethral outline throughout voiding, suggesting strongly that outlines were being determined by the external striated sphincter contracting as a whole or in part. Following urethral dilation, urethral dynamics tended to return to normal, the "spasm" urethrogram giving way to a more open urethral

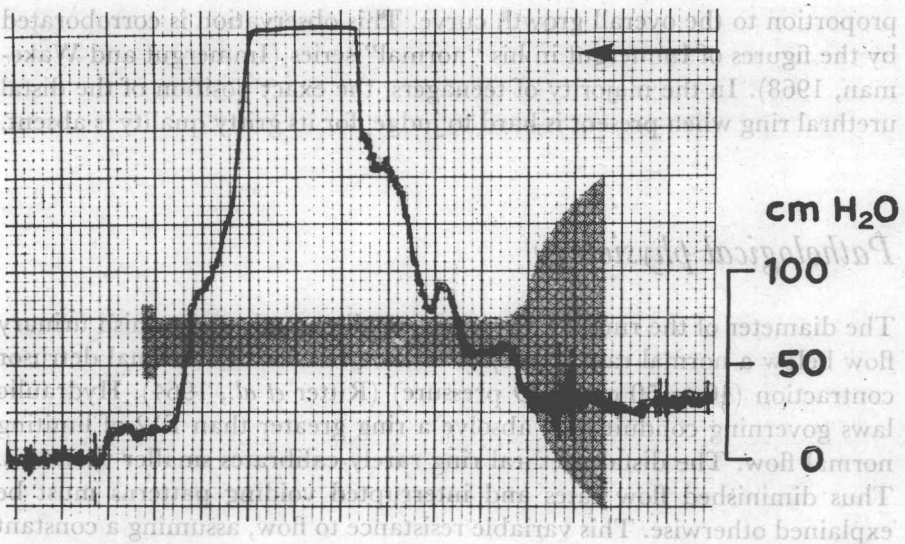

Fig. 2. Profile of urethral pressure recorded as catheter is smoothly drawn from bladder cavity to external meatus. Exceedingly high (over 225 cm H_2O) pressure in midurethral segment where the striated muscle is maximally effective. From Tanagho *et al.* (1971).

outline. Tanagho, too, considered the distal urethral ring to be only indirectly obstructive by inducing spasm of the striated external sphincter.

Similar pressure flow studies reported recently by Hendry *et al.* (1973), suggested to them that "the external striated muscle sphincter is highly active in children, and it is of interest that the girls who showed improvement after dilation were those who tended to have high urethral pressures, high bladder pressures, and low bladder capacities".

The probable mechanism for increased susceptibility to lower tract infection becomes clearer through cine studies of girls with difficult urination. The bladder urine passes into the midurethra, then regurgitates back into the bladder, turbulent flow increasing its scouring action on urethral bacteria. These are then trapped in the residual urine of 5 to 15 cc. Hinman (1966) and Mayo and Hinman (1973) logically theorized on this possibility. Following urethral dilation, flow dynamics approximate normal, turbulent flow being converted to something closer to laminar flow with its minimal regurgitative effects.

Finally, though sphincter spasm is probably not constant, it produces obstructive changes within the bladder as suggested by the increased voiding pressures measured in children with a symptomatic distal urethral ring, the presence at the least of mild trabeculation in 25 to 50 per cent of these youngsters, and a 30 per cent reversal of vesicoureteral