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# The spectrum of genital median raphe anomalies among infants undergoing ritual circumcision

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Received 3 July 2012; accepted 28 November 2012  
Available online 18 January 2013

## KEYWORDS

Genital;  
Raphe;  
Anomalies;  
Circumcision

**Abstract** *Objective:* To evaluate the extent of genital median raphe (GMR) anomalies and their relation to other genitourinary anomalies, as well as the impact on performing ritual circumcision.

*Patients and methods:* This prospective study was designed to collect data from neonates coming for ritual circumcision in order to detect any associated congenital anomalies in their genitalia, particularly in the genital raphe. 2880 babies aged from 1 day to 7 weeks were examined, from 2006 to 2011. All doubtful cases were reevaluated and cases with GMR anomalies were investigated for detection of other congenital anomalies and enrolled in the study.

*Results:* 57 cases of GMR anomalies were detected with an overall incidence of 2%; 18 of them had hypospadias, 5 had renal anomalies and 3 had limb anomalies. Circumcision was postponed in 37 cases where further investigations were done, but routine circumcision was carried out in the remaining 20.

*Conclusion:* It is crucial to examine every baby coming for circumcision to detect obvious or hidden congenital genital anomalies. Congenital anomalies of GMR are not so rare as thought, and some of these anomalies may necessitate surgical correction, and are commonly accompanied by urinary tract anomalies.

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## Introduction

The anatomy of the male genitalia varies greatly between individuals, and it is imperative for physicians to recognize

the normal spectrum and be able to identify any abnormalities, especially when reassuring anxious patients. The developmental problems of genital skin may manifest only at the first detailed inspection of a baby's genitalia during routine circumcision [1], which is done for almost all the boys in our locality at the age of 1 day–7 weeks.

Raphe means the line of union of the two halves of various symmetrical parts, and the term median raphe refers to the

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perineal raphe, which is also known as the median raphe of perineum. This line starts just anterior to the anus and extends through the scrotum, continuing on the ventral surface of the penis. In various areas the raphe takes on the name of the anatomic location of the fused skin (i.e. perineal raphe, scrotal raphe, and penile raphe also known as genital median raphe, GMR), it is usually darker in color than the surrounding skin, generally deep pink or brown [2].

GMR is the result of a fetal developmental phenomenon whereby the scrotum (the developmental equivalent of the labia in females) and penis close toward the midline and fuse, and this could explain the incidence of other genitourinary anomalies, especially hypospadias [3].

Median raphe anomalies are not considered common and are rarely taken into consideration by physicians, and can thus pass unnoticed [2]. The aim of this study was to evaluate the extent of genital median raphe (GMR) anomalies and their relation to other genitourinary anomalies, as well as the impact on performing ritual circumcision.

## Patients and methods

A total of 2880 Egyptian babies aged from 1 day to 7 weeks who presented to our pediatric surgery department for ritual circumcision were examined during a period of 6 years, from 2006 to 2011, for detection of any hidden associated genitourinary anomalies. At the end of this period, the records of 57 patients diagnosed as having GMR anomalies were reevaluated and analyzed. All these cases had been examined thoroughly, documented and investigated using appropriate tools, mainly ultrasound, ascending and voiding cystourethrogram, and MRI, to detect any other associated congenital anomalies, especially urogenital, anal or bony anomalies.

Written informed consent was obtained from parents to access and publish the patients' records and photos of the lesions, as well as the agreement of the local scientific committee at Al Azher University.

## Results

There were 57 cases of GMR anomalies detected giving an overall incidence of 2% (Table 1). The spectrum of these anomalies was very wide: hyperpigmentation in 12 cases (21%; Fig. 1), raphe deviation to one side (Fig. 2) and prominent raphe (Fig. 3) in 8 cases each (14%), short contracted raphe in 6 (10%), raphe bifurcation in 5 (8.7%) and widened raphe in 4 cases (7%) (Fig. 4), absent raphe in 3 (5.2%; Fig. 5), and also skin ridge in 3 cases (Fig. 6), raphe hypopigmentation, beaded median raphe (Fig. 7) and bucket handle raphe in 2 cases each (3.5%) (Fig. 8), and median raphe cyst and pearly penile papules (Fig. 9) in 1 case each (1.7%).

In 18 cases there were different grades of hypospadias (32%), and of these there was prominent raphe in 5 (Fig. 10), bifurcated raphe in another 5 (all cases of bifurcated raphe had hypospadias) (Figs. 11 and 12), deviation of raphe in 4 cases (Figs. 4, 13 and 14) cases of hypospadias had hyperpigmentation (Fig. 1).

Investigations of the 57 cases by ultrasound for recognition of any other associated anomalies revealed 3 cases with different renal abnormalities ranging from simple renal cyst to pelviureteric junction obstruction, 2 cases of low imperforate anus, and 3 cases of limb anomalies: absent radius and polydactyl (Table 2).

In 28 out of 57 cases of GMR the circumcision was postponed because of the presence of hypospadias in 18 cases (Fig. 14), short contracted raphe resulting in webbed penis in 6 cases, and widened raphe and skin ridge in 2 cases each.

## Discussion

Almost all Egyptian neonates undergo routine circumcision, mainly on a religious basis. There is no exact record of the prevalence of male circumcision in Egypt, but it is considered as one of the countries that have a rate of over 80% of the population [4].

**Table 1** 57 Cases of GMR anomalies among 2880 examined babies.

Anomalies	N	% Of 57 cases	% Incidence of total (2880)	Per 1000
Hyperpigmentation	12	21	0.4	4
Raphe deviation to one side	8	14	0.27	3
Prominent median raphe	8	14	0.27	3
Short raphe leads to webbed penis	6	10	0.2	2
Bifurcation	5	8.7	0.17	1.7
Wide median raphe	4	7	0.13	1.3
Skin ridge	3	5.2	0.1	1
Absent raphe	3	5.2	0.1	1
Hypopigmentation	2	3.5	0.1	1
Beaded median raphe	2	3.5	0.07	0.7
Bucket handle	2	3.5	0.07	0.7
Median raphe cyst	1	1.7	0.03	0.3
Pearly penile papules	1	1.7	0.03	0.3
Total	57		2%	20

<sup>a</sup> Right.

<sup>b</sup> Left.

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