Radiological Assessment of Anorectal Malformations; The Role of Transperineal and Infracoccgeal Sonography in Male Type Imperforate Anus

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ABSTRACT:

BACKGROUND:

Anorectal malformation is a complex spectrum of anomalies. There are different types of anorectal malformation. The distinction can usually be made on the basis of clinical data regarding the presence or absence of a visible perineal opening or passage of meconium through the vagina or urethra.

OBJECTIVE:

To assess the validity of ultrasonography in detection of the exact anomaly in male type imperforate anus, in order to help plan the proper surgical approach.

PATIENTS AND METHODS:

A prospective study carried out between December 2017 and December 2018, at pediatric surgery department in Central Teaching Hospital in Baghdad, Iraq. Nineteen patients with imperforate anus were enrolled in this study (all were males), their age ranged between 1 day and 6 months. All patients were sent for trans-perineal ultrasound and infracoccgeal ultrasound, which were performed by using a high-resolution 11 MHz linear array transducer, in which the rectal pouch-perineal distance(P-P distance), presence or absence of recto-urinary fistula along with its précised location and the relation of the blind rectal-pouch to the pubo-rectalis muscle were obtained for a proper classification of the type of imperforate anus.US findings were compared to the operative findings which were the gold standard.

RESULTS:

The rectal pouch-perineal distance(P-P) was identified using US in all patients with a mean (27.3211) mm and SD +\- (14.78631) mm. The puborectalis muscle was identified as a hypoechoic U-shaped band in all patients along with its relation to the blind rectal-pouch. There is a statistically high significant relationship between the US and the operative findings regarding the presence and precise location of the internal fistula (P-value=0.0001).US findings showed a high sensitivity of 92.8%, specificity of 100% and accuracy of 94.7%.

CONCLUSION:

U\S whether in (Transperineal \ Infracoccygeal) approach is highly accurate, it provides an excellent imaging modality with high sensitivity and specificity in the detection of the internal fistula. U\S measurements are independent of age.

KEYWORDS: Anorectal malformation, males, Transperineal & Infracoccygeal ultrasound

INTRODUCTION:

Anorectal malformations represents a wide spectrum of congenital defects of the anus, anal canal and rectum [1,2]. The urorectal septum normally separates the cloaca into urogenital and hindgut spaces, if this fails to occur normally, links between the two spaces can occur, possibly as a result of interrupted blood supply during development, imperforate anus may also occur if the cloacal membrane does not break down [3].

The terms 'low,' 'intermediate,' and 'high' are arbitrary and not useful in current therapeutic or prognostic terminology, a therapeutic and prognostically oriented classification is depicted [4] as in table (1).

Central Child Teaching Hospital/ Baghdad, Iraq.

Table (1) Anatomical classification of Infants with Anorectal Malformations from Ashcraft's Pediatric Surgery,2014.[4]

Male	Female
• Rectoperineal fistula • Rectourethral bulbar fistula • Rectourethral prostatic fistula • Rectobladder neck fistula • Imperforate anus without fistula • Rectal atresia/rectal stenosis	• Rectoperineal fistula • Rectovestibular fistula • Cloaca • Complex malformations • Imperforate anus without fistula • Rectal atresia/rectal stenosis

AIM OF THE STUDY:

This study was conducted to assess the validity of ultrasonography in detection of the exact anomaly in male type imperforate anus, in order to help plan the proper surgical approach.

PATIENTS AND METHODS:

Study design and setting: A prospective study carried out between December 2017 and December 2018 at the department of pediatric surgery in Central Child Teaching Hospital in Baghdad, Iraq.

The radiological assessments were done at the department of radiology in Central Child Teaching Hospital in Baghdad by the same radiologist & the technician.

Patients: (26) male patients were examined, but only (19) patients were enrolled in this study, their age ranged between 1 day and 6 months. The inclusion criteria were any male patient with imperforate anus in which meconium was noted in the urine and imperforate anus with no fistula, regardless of the duration of illness since the diagnosis was made. (7) patients were excluded from this study for the following reasons as mentioned in front of each category as it shows in table (2).

Table (2) Causes for patient's exclusion from the study

Causes for exclusion	Number Of patients
Males with recto-perineal fistula	4
Males died from other causes before completing this study	1
Males who were enrolled in this study, but didn't complete PSARP	2
Total	7

Data collection: All patients with imperforate anus were admitted to pediatric surgical ward in Central Child Teaching Hospital in Baghdad. A detailed history and clinical examination was obtained according to a pre-designed questionnaire with a special emphasis on age, presence of perineal fistula, bucket handle malformation, anterior displaced anus, presence of meconium in the urine, whether the anal dimple was prominent or barely visible, whether the development of the sacrum was well, poor or short and deformed ,if the midline groove was prominent or poorly developed, if the perineum was flat or well developed, any associated anomalies, family history of imperforate anus. Radiological assessment for all (19) candidates enrolled in this study were done by transperineal & infracoccgeal ultrasound, in which the rectal pouch-perineal distance(P-P) measured in millimeters, presence or absence of recto-urinary fistula along with its précised location and length and the relation of the blind rectal-pouch to the puborectalis muscle was obtained.

Transperineal ultrasound was performed by using a high-resolution 11 MHz linear array transducer (Vulson EL, GE. Healthcare ultrasonography machine,2015) that executed by the same radiologist. All (19) candidates were examined in a modified lithotomy position without specific preparation. All sonographic measurements were accomplished in the resting state while the child was calm & comfortable, meanwhile the mother or care giver was holding a pacifier or a rattle aimed for child distraction. A gel material (Konix ultrasound gel) was applied to the perineum. Care was taken so no pressure was applied to the perineum during examination. Scanning was implemented in mid-sagittal plane through the perineum, as in figure (1).

Figure (1) TPU scanning in mid-sagittal plane.

The bladder, the urethra and distal rectal pouch were identified as in figures (2) and (3), while in the infracoccygeal approach, all the patients were examined in a supine position with both legs pulled up to the chest.

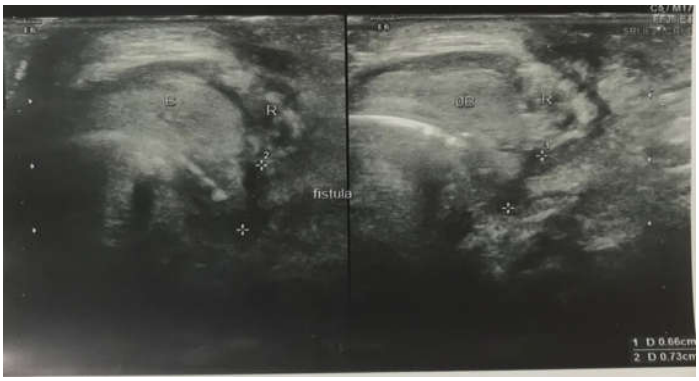

Figure (2) TPU shows the blind rectal pouch with rectourethal fistula.

Figure (3) TPU shows the blind rectal pouch with recto-bladder neck fistula..

Scanning was implemented in the coronal plane as in figure (4), through the infra coccygeal region where the puborectalis muscle was identified and its relation to rectal pouch was determined, as in figure (5).

Figure (4) Infracoccgeal U\S scanning in coronal plane.

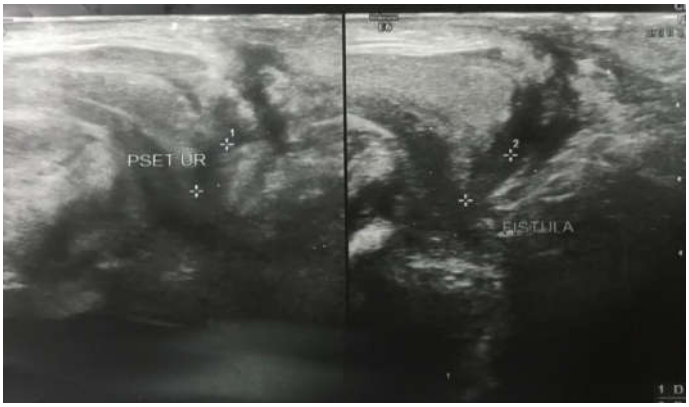

Figure (5) Infracoccgeal U\S shows the distal rectal pouch bellow puborecatalis muscle..

All (19) patients enrolled in this study were scheduled for definitive repair (PSARP). At time of operative repair all patients were admitted to the surgical ward where baseline investigations were obtained, blood preparation was done, bowel preparation was accomplished, prophylactic antibiotics were given. At the operative room all patients were placed in prone position with pelvis slightly elevated once general anesthesia was attained and Foley's catheter was inserted, painting the operative field from the lower back to the mid-thigh was accomplished, defining the site for the external anal sphincter was performed using Pena stimulator, then the operative procedure (PSARP) was completed while using a sterilized ruler for measurement purposes in which the rectal-pouch-perineal (P-P) distance was measured, along with outlining the presence or the absence of recto-urinary fistula , and its précised location.

Ethical consideration: Approval from both pediatric surgery and Radiology departments in Central Child Teaching Hospital in Baghdad and the Iraqi Board of Medical Specialty was taken before the start of gathering patients. Consent was taken from each patient after clearly explaining the aim of the study and the benefit from the role of radiological assessment in their status. The Patient identity and privacy were kept classified during the assessment.

Statistical analysis: The data were analyzed using the SPSS (SPSS Inc., Chicago, IL, USA) version 22. Chi-square test was used to find the significance of differences in categorical parameters.

RESULTS:

Nineteen male patients were enrolled in this study (13) were neonates and (6) were infants, their age reneges between (1-180) days, with a mean (31.2105) days and SD +\- (50.57841) days. Table [3] shows the baseline findings in our study.

Table [3]: base line patient's data

	Characteristic	Minimum	Maximum	Mean +/- SD
1	Age (mean)	1 day	180 days	31.2105 +/- 50.57841 days
2	U\S P-P distance	12.80 mm	64.00 mm	(27.3211) SD +\- (14.78631) mm.
3	Operative P-P distance	15.00 mm	60.00 mm	(29.4737) SD+\- (13.52926) mm.

and operative findings is greater than 15mm which is considered the cut off point for the high type ARM .

Distribution of U/S and operative findings of the presence and location of internal fistula is shown in figure [6]. Of the 19 patients enrolled in the study, U/S detect 13 patients (68.4%) had internal fistula, of those 10 patients (52.6%) had rectouretheral and 3 patients (15.8%) had rectobladder -neck fistulas and only 6 patients (31.6%) had no fistula detected. The operative procedure detects 14 patients (73.7%) of the total 19 patients enrolled with internal fistula, 11 patients (57.9%) had rectouretheral and 3 patients (15.8%) had rectobladder- neck fistula and in only 5 patients (26.3%) no fistula was detected figure (7).

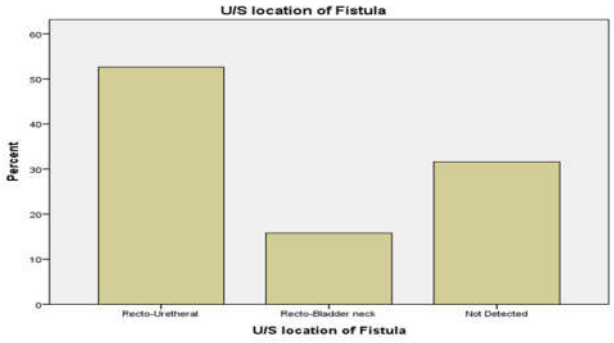

Figure (6) Distribution of the U\S findings regarding the presence and location internal fistula in the studied patients.

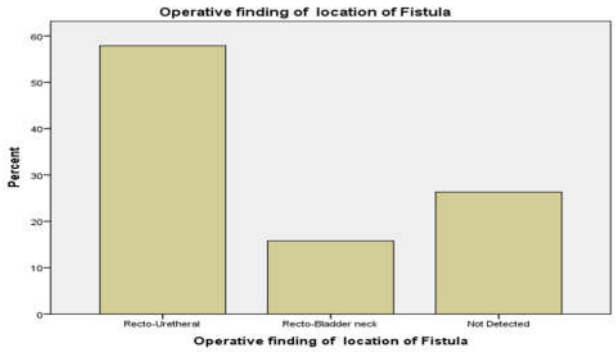

Figure (7) Distribution of the operative findings regarding the presence and location internal fistula in the studied patients.

The value of U/S in detection of the relation between the rectal pouch and the puborectalis muscle was studied as shown in figure [8]. Of the 19 patients enrolled 11 patients (57.9%) had the rectal pouch below and 8 patients (42.1%) above the puborectalis muscle. in which rectal pouch usually ends 1-2 cm higher from the perineal skin and this makes it below the puborectalis muscle these are considered to be a low type IA

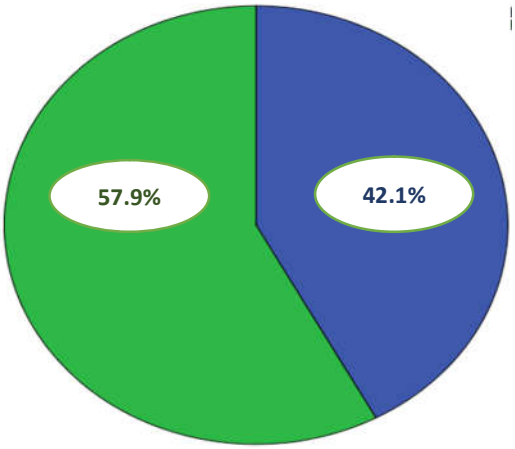

Figure (8) Distribution of the U\S findings regarding the relation of the rectal pouch to puborectalis muscle in the studied patients.

Different variables relationships and P-value was measured as shown in table [4]. No significant relationship was found between the age, U/S P-P distance, and Operative P-P distance, sensitivity in finding the internal fistula by U/S, and Operative technique.

We found a statistically high significant relationship between the sensitivity of U/S and Operative technique in detection and accurate location finding of the internal fistula (P-value = 0.0001) for both. The U/S in both transperineal and/or infracoccygeal scanning had an overall 92.8% sensitivity, 100% specificity and 94.7% accuracy.

Table [4]: different variables and their P-value in our study

	Relationships	P-value
1	Relation between age and U/S P-P distance finding	0.275
2	Relation between U/S and Operative P-P distance	0.308
4	Relation between the sensitivity of U/S and Operative technique in finding of internal fistula	**0.0001**
6??	Relation between the accuracy of precise detection of location of internal fistula between U/S and Operative technique	**0.0001**

DISCUSSION:

The final diagnosis of Anorectal anomalies, including the type of anomaly was made based on imaging and surgical findings according to the international classification of anorectal anomalies [5]. The distinction can usually be made based on clinical data regarding the presence or absence of a visible perinea opening or passage of meconium through the vagina or urethra [6]. The role of a radiologist is to evaluate and classify ARM and associated anomalies with available imaging modalities to help the clinician to decide on therapeutic strategy [7]. We have found a mean age of (31.2105) days, similar to the finding of Kim et al [8] and Haper et al [9] in which the mean age was (1.1) month, and (30) days, respectively. However, it goes against Han et al [10] with a mean age of (6) days. This difference because the last chose to take on a small sample through which neonates were selected exclusively. Conversely, because infants were included in our study.

We have found that there is no statistically significant relationship between the patient-age and U\S finding regarding the P-P distance, (P-value = 0.275). this appears to be similar to Hosokawa et al [11] findings (P-value = 0.26). This is probably because the P-P distance only appears longer before completing 24 hour of life, however after 24 hours and once the gas or meconium have reached the blind distal rectal pouch, the distance becomes in dependable of the age. However, unless something causes an increase in the intra-abdominal pressure like straining or crying for instance, which in this case gives false positive shorter distance, it is constant and non-age related after the post-natal day one of U\S examination.

The US P-P distance findings was similar to operative findings in this study in which the mean was (27.3211) mm with SD +\- (14.78631) mm, (39.5789) mm SD +\- (13.12045) mm and (29.4737) mm, SD+\- (13.52926) mm respectively. The US findings were compatible with the finding in Haber et al [9], Jardosh et al [12] and choi et al [13] which showed a mean of 24 ± 6 mm, 20.4 +\- 4.7 and 18.2 mm, however it was mismatched with Han et al [10] and Hosokawa et al [11] which was 12.2 mm and 8.3 +\-2.9 respectively.These findings can be explained by the dominate presence of imperforate anus with recto-urinary fistula (urethral and vesicle) in this study and since the more complex the defects the higher the rectum terminates from the perineal skin, consequently the higher the P-P distance becomes ,and hence the mean for the P-P distance in US , and operative findings is greater than 15mm which is considered the cut off point for the high type ARM . We have found that there is no statistically significant relationship between the U\S P-P distance and the operative P-P distance (P-value = 0.308). It is parallel with Niedzielski et al [14] (P-value>0.001) however, this goes in contradiction with the findings of Haber et al [9], Jardosh et al [12] and Alehossein et al [15], in which the (P-value < 0.001) for all three of them in which there is a significant difference between the U/S P-P distance and the operative P-P distance. This might be due to the easy sonographic delineation of the distal rectal pouch as a blind end hypoechoic structure containing either meconium or gas, and since the neonate or the infant was kept calm during the examination session, the precise distance can be estimated, however since the results came back with some overlap with operative findings

in which only few millimeters difference representing narrow margin for error so no significant difference between the two variables was estimated. Furthermore, this is could be related to the unfamiliarity of the radiologist with this simple, noninvasive but rather innovative sonographic approach.

We have found that (57.9%) of the studied patients had the rectal pouch below the puborectalis muscle and in (42.1%) the rectal pouch was above it. These findings correlate with Han et al [10] in which 71.5% of the cases the rectal pouch was above the puborectalis muscle and 28.5% below it. This distribution can be clarified by the presence of a high percentage of Imperforate anus(IA) with recto-urethral (bulbar) fistula and IA without fistula that was detected in this study, in which rectal pouch usually ends 1-2 cm higher from the perineal skin and this makes it below the puborectalis muscle these are considered to be a low type IA and will have a relatively good prognosis regarding the continence in the future, while the more complex forms of IA with recto-urethral (prostatic) fistula or recto-bladder neck fistula ,the rectum tends to terminate higher, above the puborectalis muscle which is usually poorly developed and thus, these patients characterize by having a poor prognosis.

Regarding the U\S distribution for presence and location of internal fistula in the studied patients, we have found that the US findings correlate with operative findings. We found these results to be parallel with Choi et al [9], Jardosh et al [12] which detected internal fistulae in 60.8% , 66.66% respectively, Jardosh et al [13] also described that out of 23 male patient a recto-urethral fistula was found in 8 (34.7%), and Oguma et al [64] where(74%) had internal fistula while in (26%) no fistula was detected.

However, our findings appear to be nonparallel with Sarkar et al [16] and Kim et al [17] where (84%) had internal fistula, while in (16%) it was not detected (92%) had recto-urethral fistula and (7.7%) had rectobladder neck fistula respectively.

The relatively high proportion of imperforate anus without fistula in our study might be due to the association with Down syndrome which was priory identified in 4 out of 5 of the cases having IA without fistula. As for the presence and distribution of the internal fistula in the studied patients, it can be explained by the dominate presence recto-urethral fistula which is considered to be the most common type in male IA, followed by recto-bladder neck fistula in a lesser degree which tends to occur in about 10% of male IA. We have found that there is a statistically highly significant relationship between the U\S and the operative findings regarding the presence and the precise location of internal fistula (P-value = 0.0001) in which sensitivity was 92.8%, specificity 100% and accuracy 94.7%. These findings were paralleled to the findings of the studies listed in table (5). These findings can be clarified by the ability of US to image the end of the rectal pouch, regardless of whether it is impacted with meconium or air which is separated from the anteriorly located urogenital tract by fat that appears to be echogenic on US. The internal fistula however, can be identified as hypoechoic linear tract piercing through the echogenic fat plain, with the advantage of a real time US scanning this shows its superiority and the accuracy

Table (5) List of studies including their U\S findings regarding the presence and the precise location of internal fistula used for comparison to our current study.

Study	Niedzielskiet et al[14]	Hosokawa et al[18]	Choi et al[13]	Haber et al[9]	Jardosh et al[12]	Hosokawa et al[19]	Our study
P-value	< 0.001	0.012					0.0001
Sensitivity	86%			100%	87.5%	75.5%	92.8%
Specificity				86%	100%	75.5%	100%
Accuracy			85%	95%	91.3%	75.5%	94.7%

CONCLUSION:

U\S whether in (Transperineal\Infracoccygeal) approach is highly accurate, provide excellent imaging modality with high sensitivity and specificity, enables the classification of IA based the identification of the exact P-P distance, the presence of the internal fistula with its precise location and relation of distal rectal pouch to puborectalis muscle. Thus, it guides the initial surgical decision about the need for a simple anoplasty or colostomy. It also influences the surgical approach at the time of definitive repair whether merely Posterior Sagittal Ano-Recto PLasty (PSARP) or there would be a need for accompanying laparotomy or laparoscopy. U\S measurements are independent of age and can be performed in both neonate and infant with an equivalent level of precision as long the patient is over 24 hours old.

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