Original Article

Prevalence and Association of Occult Spinal Dysraphism with Anorectal Malformation

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Background: Anorectal malformation (ARM) is associated with occult spinal dysraphism (OSD). The guideline for screening for OSD in patients with ARM is still controversial.

Objective: 1) To evaluate the prevalence of OSD in each type of ARM and to elucidate which types of ARM should be screened for underlying OSD. 2) To compare the methods used in radiologic screening for OSD, such as plain lumbosacral X-ray, spinal ultrasound (U/S), and spinal magnetic resonance imaging (MRI).

Materials and Methods: The medical records of all patients with ARM treated at Siriraj Hospital between 2006 and 2016 were reviewed. Spinal dysraphism screening with lumbosacral X-ray, U/S, or MRI were performed depending on the surgeon's request. Associated urinary tract anomalies were noted. Data of the surgical treatments, outcomes, and complications of treatments were collected.

Results: Among the 204 ARM patients treated, 150 patients were screened for OSD, involving 150 lumbosacral X-ray, 65 U/S, and 39 MRI. The prevalence of OSD (n=33) in all types of ARM was 16.2%. The higher types of ARM had higher prevalence of OSD, except for vestibular fistula (20.5%). The prevalence of OSD in cloaca malformation, rectobladder neck fistula, rectoprostatic urethral fistula, rectobulbar urethral fistula, and perineal fistula were 28.6%, 33.3%, 16.7%, 12.5%, and 9.8%, respectively. Lumbosacral X-ray had a sensitivity of 51.7% and was not a good screening method. Sixteen of 65 U/S revealed OSD (24.6%). Tethered cord and syringomyelia were noted in nine and seven, respectively. Spinal MRI (n=39) revealed 27 OSD (69.2%) comprising 16 tethered cords, 12 spinal lipoma, and 10 syringomyelia. ARM associated with KUB anomalies had a statistically significant correlation with OSD. In 33 OSD, 17 patients (51.1%) were operated on. Fourteen received untethering spinal cord, while five had intradural lipoma excised.

Conclusion: Patients with ARM had a high prevalence of OSD and should be screened for. U/S is the first-line method. MRI is strongly recommended in cloaca malformation, rectobladder neck fistula, rectoprostatic urethral fistula, vestibular fistula, and all ARM associated with KUB anomalies. Other types of ARM should be screened by MRI if feasible.

Keywords: Spinal dysraphism, Tethered cord, Anorectal malformation

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Anorectal malformation (ARM) is known to be definitely associated with other congenital defects, including spinal dysraphism. Spinal dysraphism includes congenital spinal abnormalities resulting from an improper midline fusion of bony, mesenchymal, and neural structures, such as intraspinal masses, lipomyelomeningoceles, tethered cord, and occult meningocele. Patients with ARMs who have a spinal area with lumps, nevus, sinus, angioma, hypertrichosis, skin dimples, or sacral lipomas[1,2] may be suspected to have spinal dysraphism, therefore, further investigations for the diagnosis and planning of treatments should be arranged. However, if there was no evidence of spinal dysraphism, defined as occult spinal dysraphism (OSD, which comprises tethered cord, diastematomyelia, neuroenteric cyst, and lipoma[3]), the rational for performing screening tests is still considered controversial.

The reason why OSD, especially a tethered cord, should be diagnosed early, is because of the risk of a progressive neurologic deterioration of the spinal cord. A tethered cord involves a fixation of the lower end of the spinal cord (Figure 1). During growth and development, the increased traction on the

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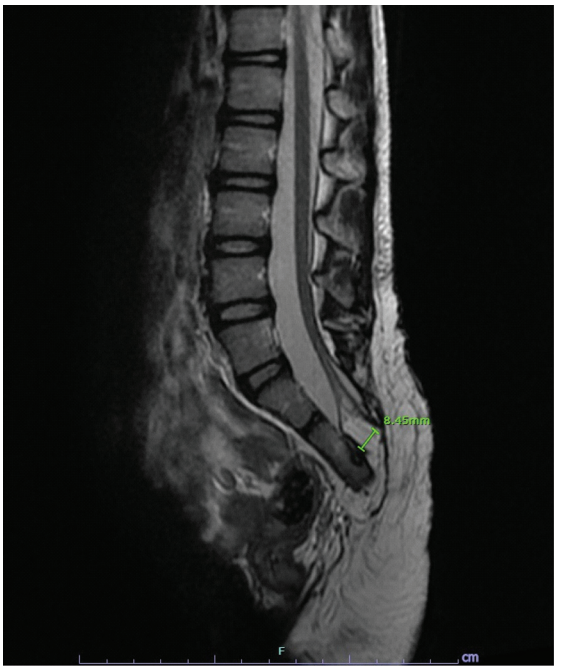

Figure 1. Sagital MRI T2W revealed incomplete development of sacral bone, spinal cord lipoma, low lying of spinal cord and tethered cord.

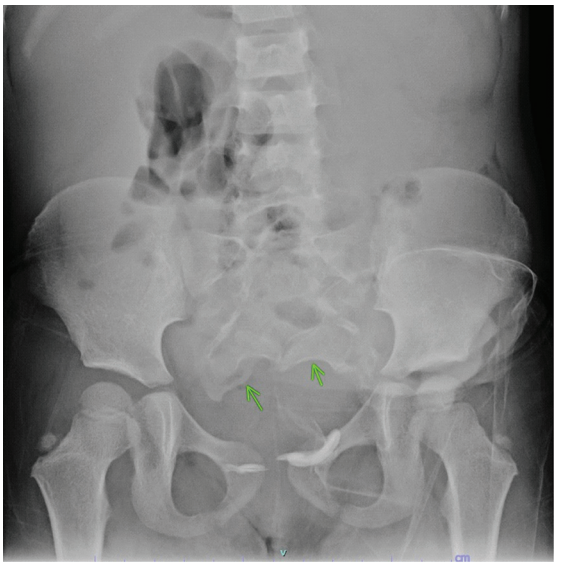

Figure 2. Plain film pelvis revealed incomplete development of sacral bone (arrow).

tethered cord could result in progressive symptoms and may display signs involving the motor-sensory, orthopedic, urinary, or bowel functions. Orthopedic symptoms usually present as weakness or atrophy, abnormal reflexes, pain in the back or legs, sensibility disorders, deformities, and gait abnormalities. Urinary involvement includes incontinence or a persistent urge due to a neurogenic bladder, while bowel symptoms could be soiling and fecal accidents. The early recognition and treatment of correctable lesions of the spinal cord in patients with ARMs may preserve important neurologic functions. Prophylactic surgical untethering in patients who have no neurological deficit would be of great benefit[1,3-6].

Each radiologic modality has some limitations. A plain lumbosacral X-ray could demonstrate spina bifida, an abnormal spinal curve, and hemivertebrae (Figure 2), but incomplete ossification in a plain lumbosacral X-ray of the neonatal spine would make it difficult to conclude on the results. High-resolution ultrasonography is considered a rapid, safe, non-invasive method of evaluation because of the associated acoustic window provided by the spina bifida and incomplete ossification of the posterior elements in infants. Moreover, it could be done at the bedside and does not require sedation. The limitations of ultrasound (U/S) are the operator dependence and an inability to identify a tethered cord in the newborn period. The advantages of spinal magnetic resonance imaging (MRI) are the lack of ionizing radiation and a superb demonstration of the anatomy and pathology, while the disadvantages include the high cost of MRI, the need for sedation, and difficulties in imaging in severe scoliosis.

Whether every ARM should be screened by spinal MRI was the principal question in the present study. Levitt et al[7] published a large series of ARM and revealed that tethered cord was found in 24% of the patients, while other researchers have revealed a prevalence of tethered cord of between 10% and 52%[4,8,9]. Some studies recommend that spinal MRI should be conducted for every case of ARM[1,4,6,9,10]. However here in the Division of Pediatric Surgery, Department of Surgery, Faculty of Medicine, Mahidol University, Thailand, the authors did not adhere to this protocol.

From previous publications, some entities of ARM demonstrated a high prevalence of OSD and tethered cord. One study reported the following findings:

1. The more complex the ARM, the higher the prevalence of tethered cord[7].

2. Plain lumbosacral X-rays that had revealed sacral hypo-development, presacral mass, or sacral hemivertebrae had a higher prevalence of OSD and tethered cord[7].

3. ARM associated with KUB anomalies, such as single kidney, ectopic kidney, hypospadias, hydro-

ureter, hydronephrosis, and dysplastic kidney, also had a higher prevalence of OSD and tethered cord[7].

Consequently, the aim of the present study was to answer the question of whether every ARM case should be screened for OSD or should only some entities of ARMs be screened, and if screening were to be applied, which method of screening would be the best investigation to perform. All patients diagnosed with ARM at the Division of Pediatric Surgery, Department of Surgery, Faculty of Medicine, Mahidol University, Thailand, were reviewed.

Objectives

1. The aim of the present study was to evaluate the prevalence of OSD in each type of ARM and to elucidate which types of ARM should be screened for underlying OSD.
2. The methods used in radiologic screening for spinal dysraphism, such as plain lumbosacral X-ray, spinal U/S, and spinal MRI, were compared.
3. The prevalence of OSD in ARM with other associated congenital anomalies, such as KUB anomalies, was evaluated for the prevalence of OSD to assess whether it was high enough to justify screening for OSD.

Materials and Methods

The present study was approved by Siriraj Institutional Review Board, COA No. Si764/2017. A retrospective review was performed of the medical records and radiologic findings of all patients with ARMs that underwent surgical management at the Division of Pediatric Surgery, Department of Surgery, Faculty of Medicine, Mahidol University, Thailand, between January 2006 and December 2016.

The exclusion criteria were 1) bladder exstrophy, 2) cloacal exstrophy, 3) inadequate follow-up period, 4) death before full neurological investigations, 5) major chromosome anomaly that involved an incapability to survive, or 6) incomplete medical records.

Krickenbeck's classification was used to classify the types of ARM. Spinal dysraphism screening with a plain lumbosacral X-ray, spinal U/S, or spinal MRI were performed in some patients depending on the surgeon's request. Any associated urinary tract anomalies were noted.

Data including the patients' basic demographic data, types of ARM, types of spinal cord anomaly, types of spinal anomaly, findings of the plain lumbosacral X-ray, spinal ultrasonography, or spinal MRI, symptoms of spinal cord abnormality, the surgical treatments, outcomes, and complications of spinal dysraphism treatments, and any associated urinary tract anomalies were collected. All the data were processed through the SPSS Statistics software and statistical analysis was carried out.

Table 1. Prevalence of spinal dysraphism in each type of ARM

Types of anorectal malformation	No. of patients	No. of spinal dysraphisms; n (%)
Cloaca malformation	21	6 (28.6)
Rectovesical fistula	15	5 (33.3)
Rectourethral (prostatic fistula)	24	4 (16.7)
Rectourethral (bulbar fistula)	16	2 (12.5)
Vestibular fistula	39	8 (20.5)
Perineal (cutaneous) fistula	51	5 (9.8)
ARM with no fistula	38	3 (7.9)
Total	204	33 (16.1)

ARM=anorectal malformation

Results

In between 2006 and 2016, there were 204 patients with ARM (male 115, female 89) treated in the Division of Pediatric Surgery, Department of Surgery, Siriraj Hospital, Mahidol University, Thailand.

Of these 204 ARM patients, plain lumbosacral X-ray was done in 150 patients screened for spinal anomalies, while 84 patients were screened for spinal cord anomalies, involving 65 U/S or 39 spinal MRI tests. Some patients were screened using both U/S and MRI. The other patients who did not receive either U/S or MRI were continuously assessed for neurological abnormalities in their long-term follow-ups in the outpatient clinic.

OSD was found in 33 ARM patients and the overall prevalence of OSD in all types of ARM patients was 16.2%.

Prevalence of spinal dysraphism in each type of ARM

The different types of ARM have a different prevalence of spinal dysraphism. The more complex the ARM, the higher the prevalence of spinal dysraphism. Surprisingly, vestibular fistula had a high prevalence of spinal dysraphism (20.5%). The prevalence of spinal dysraphism in cloaca malformation, rectovesical fistula, rectoprostatic urethral fistula, rectobulbar urethral fistula, and perineal fistula were 28.6%, 33.3%, 16.7%, 12.5%, and 9.8%, respectively. The prevalence of spinal dysraphism in each type of ARM is demonstrated in Table 1.

Table 2. Symptoms might be related to spinal dysraphism in patients with anorectal malformation

Symptoms	No. of patients
Motor symptoms	3
Scoliosis	1
Orthopedic, abnormal gait, foot deformity	2
Urinary incontinence/disturbances	4
Fecal incontinence	8

Table 3. Abnormalities of the spine detected by plain spinal X-ray (n=29/150)

Spinal abnormality	No. of patients
Sacral hypodevelopment	16
Sacral hemivertebrae	4
Lumbar hemivertebrae	3
Spina bifida	2
Thoracic anomalies	7
Scoliosis	7

Overall, 13 of the 204 patients (6.4%) had symptoms that might be related to OSD. The most common symptoms included motor symptoms, scoliosis, orthopedic gait abnormalities, urinary incontinence, and fecal incontinence, as shown in Table 2. Some patients had more than one symptom. It was hard to conclude for fecal incontinence whether it was caused by spinal dysraphism or ARMs.

Spinal screening with a plain X-ray of the spine

Most of the patients with ARM were screened for spinal anomalies with a plain X-ray of the lumbosacral spine (n=150) and some received a thoracic spinal X-ray. The abnormalities of the spine detected by a plain X-ray of the spine are revealed in Table 3. Vertebral spine anomalies were detected by a plain X-ray of the spine in only 29 patients (19%). Plain spinal X-ray could detect some spinal anomalies with a sensitivity of 51.7% (95% CI 34.4 to 68.6), specificity of 88.4% (95% CI 81.5 to 93), accuracy of 81.3% (95% CI 74.3 to 86.3), and positive predictive value of 51.7%. Thus, a plain spinal X-ray is not considered a good modality for screening spinal anomalies in patients with ARM and may not be able to identify all the spinal cord lesions.

Spinal cord and spinal anomaly screening with spinal ultrasound

U/S can identify either spinal cord anomalies or spinal anomalies. Among the 65 patients that underwent U/S, abnormal ultrasonographic findings were found in 16 patients (24.6%). The abnormal findings detected by ultrasonography (n=16/65) are shown in Table 4.

Table 4. Abnormal findings detected by ultrasonography (n=16/65)

Ultrasonographic findings	No. of patients; n (%)
Spinal cord anomaly	
Tethered spinal cord	9 (13.8)
Syringomyelia	7 (10.8)
Filar cyst	3 (4.6)
Thick filum terminale without tethered cord	1 (1.5)
Spinal lipoma	1 (1.5)
Spine anomaly	
Spina bifida	1 (1.5)
Sacral hemivertebrae	1 (1.5)

Spinal cord and spinal anomaly screening with spinal MRI

MRI could identify either spinal cord anomalies or spinal anomalies. Among the 39 patients who underwent spinal MRI, abnormal MRI findings were found in 27 patients (69.2%). Some patients had two or more spinal cord and spinal anomalies. The pathologic findings detected by MRI (n=27/39) are demonstrated in Table 5.

Patients with ARM were screened for KUB abnormalities, and these were done in 185 patients.

Urinary tract abnormalities were found in 65 (31%) patients with ARM (n=204). KUB abnormalities comprised of 44 (23.8%) cases of hydronephrosis, 26 (14.1%) vesicoureteric reflux, 18 (9.7%) hydroureter, 14 (7.6%) renal agenesis, seven (3.8%) neurogenic bladder, five (2.7%) multicystic dysplastic kidney, four (2.1%) crossed fused renal ectopia, four (2.1%) bladder diverticulum, three (1.6%) ureteropelvic junction (UPJ) obstruction, three (1.6%) horseshoe kidney, three (1.6%) ectopic ureter, two (1.1%) ureterovesical junction (UVJ) obstruction, two (1.1%) renal cysts, and one each of a posterior urethral valve and double collecting system. Patients with ARM associated with KUB anomalies had a statistically significant correlation with OSD according to the Pearson chi-square test (p=0.001).

Treatments for occult spinal dysraphism

In the present study, 33 (16.1%) of the 204

Table 5. Pathologic findings detected by MRI (n=27/39)

MRI findings	No. of patients; n (%)
Spinal cord anomaly	
Tethered spinal cord	16 (41.0)
Spinal lipoma	12 (30.8)
Syringomyelia	10 (25.6)
Lipomyelomeningocele	2 (5.1)
Filar cyst/perineural cyst	2 (5.1)
Lipomeningocele	1 (2.6)
Thickened fatty filum with mild low lying cord	1 (2.6)
Spine anomaly	
Sacral hypodevelopment	12 (30.8)
Spina bifida	8 (20.5)
Sacral hemivertebrae	2 (5.1)
Butterfly vertebrae	2 (5.1)
Partial/unfused vertebrae	2 (5.1)
Coccyx agenesis	1 (2.6)
Presacral tumor	1 (2.6)

MRI= magnetic resonance imaging

Table 6. Surgical treatments of spinal dysraphism (n=17)

Surgical treatment	No. of patients; n (%)
Untethering spinal cord	14 (82.4)
Excision intradural lipoma	5 (29.4)
Repair meningocele	1 (5.9)
Excision presacral mass	1 (5.9)
Syringosubarachnoid shunt	1 (5.9)

Table 7. Outcomes of surgical treatments in all operated cases (n=17)

Outcomes	Preoperative symptoms	Improved	Not improved
Motor symptoms	3	3	0
Scoliosis	1	1	0
Orthopedic abnormal gait	2	2	0
Urinary incontinence	3	2	1
Fecal incontinence	3	2	1

patients with ARM had a radiologic diagnosis of spinal dysraphism. Thirteen patients (6.4%) had symptoms that might be related to OSD including motor symptoms, scoliosis, orthopedic gait abnormalities, urinary incontinence, and fecal incontinence. Excluding orthopedic problems, nine patients had symptoms of neuromotor deterioration or urinary or fecal incontinence.

Spinal cord operations were conducted in 17 patients. Among the nine symptomatic neuromotor deterioration or urinary or fecal incontinence, eight patients were operated on, except one with a symptom of fecal incontinence. Among the other 24 spinal dysraphism patients, prophylactic operations were conducted in nine patients. The remaining 15 asymptomatic patients were closely followed up for spinal cord abnormalities symptoms at the outpatient clinic of Siriraj Hospital. The operative treatments depended on the spinal abnormalities and the preoperative clinical symptoms, which are listed in Table 6. Most of the operations were for untethering the spinal cord (82.4%) and excision of intradural lipoma (29.4%).

In the symptomatic group, most patients had more than one symptom. Six of the eight symptomatic patients that underwent an operation showed improvements in their symptoms postoperatively, whereas two patients did not show improvements for urinary and fecal incontinence postoperatively. In the asymptomatic group operated on for prophylactic reasons, no patient showed a deterioration in their neurological signs and symptoms postoperatively. The outcomes of the surgical treatments in all the operated cases are revealed in Table 7.

Of the 17 patients that underwent surgical treatments, five patients (29.4%) had complications. Here, one patient with CSF leakage developed pseudomeningocele on the twentieth day post-operatively and a repair of the pseudomeningocele was further performed. The other complications, which were one persistent or recurrence of the tethering cord, one wound dehiscence, one wound infection, and one hydrocephalus, showed improvements without the need for a second operation.

The minimum age of patients suitable for screening using spinal ultrasonography or spinal MRI is another controversial issue. For patients screened before 24 months of age, it was possible to detect asymptomatic spinal dysraphism in 70 cases (92.1%), which was higher than for the five patients (62.5%) screened after 24 months of age. Screening the spinal cord before 24 months of age allowed detection in patients before they had developed neuromotor deterioration or urinary or fecal incontinence symptoms (7.9%), which was a much higher detection rate than from screening the spinal cord in patients older than 24 months of age (37.5%), with a statistically significant difference ($p<0.05$). Comparisons between the age of screening using spinal ultrasonography or spinal MRI and the

Table 8. Comparison between age of screening using spinal ultrasonography and/or spinal MRI and ability to detect asymptomatic spinal dysraphism

Age of screening	Asymptomatic; n (%)	Symptomatic*; n (%)
Before 24 months	70 (92.1)	6 (7.9)
After 24 months	5 (62.5)	3 (37.5)

* Neuromotor deterioration or urinary/fecal incontinence

ability to detect asymptomatic spinal dysraphism are revealed in Table 8.

Discussion

Here, spinal dysraphism included those congenital spinal abnormalities resulting from an improper midline fusion of the bony, mesenchymal, and neural structures, such as intraspinal masses, lipomyelomeningoceles, tethered cord, and occult meningocele. ARMs were definitely associated with other congenital defects, including spinal dysraphism[7]. Levitt et al[7] published a large series of data on ARM and revealed that tethered cord was found in 24% of patients with ARM, but other researchers have reported a prevalence of tethered cord between 10% to 52%[4,6,8,9,11]. In the present series, the prevalence of OSD in patients with ARM was 16.2%. Patients with ARM whose spinal area has lumps, nevus, sinus, angioma, hypertrichosis, skin dimples, or sacral lipomas[1,2] are usually suspected to have spinal dysraphism, and therefore, further investigations for the diagnosis and planning of treatment would be arranged. However, patients with ARMs who have no evidence of spinal dysraphism, defined as OSD, present a greater challenge for a pediatric surgeon to diagnosis this condition. If OSD was left unattended, deterioration involving neurologic deficits would progress.

OSD comprises tethered cord, diastematomyelia, neuroenteric cyst, and spinal lipoma[3]. Tethered cord involves a fixation of low-lying conus medullaris to the sacral spine. During growth and development, the increased traction on the tethered cord could result in progressive symptoms and signs involving the motor-sensory, orthopedic, urinary, and bowel functions. The reason that OSD, especially a tethered cord, should be diagnosed early is to halt the progressive neurologic deterioration of the spinal cord. Orthopedic symptoms present as a weakness or atrophy, abnormal reflexes, a pain in the back or legs, sensibility disorders, deformities, and gait abnormalities. Urinary involvement includes incontinence or a persistent urge due to a neurogenic bladder, while bowel symptoms include soiling and fecal accidents. The early recognition and treatment of correctable lesions of the spinal cord in patients with ARMs may preserve important neurologic functioning. Prophylactic surgical untethering in patients with a tethered cord who had no neurological deficit would be of great benefit[1,3-6].

The aim of the present study was to evaluate the prevalence of OSD in each type of ARM and to elucidate which types of ARM should be screened for underlying OSD. From a large series study of patients with ARM associated with tethered cord, it was reported that the more complex the ARM, the greater the prevalence of tethered cord[7]. In the present study series, the different types of ARM had different prevalence of spinal dysraphism. The more complex the ARM, the greater the prevalence of spinal dysraphism, except for vestibular fistula, which also had a high prevalence of spinal dysraphism (20.5%). The prevalence of spinal dysraphism in cloaca malformation, rectovesical fistula, rectoprostatic urethral fistula, rectobulbar urethral fistula, and perineal fistula were 28.6%, 33.3%, 16.7%, 12.5%, and 9.8%, respectively.

ARM associated with KUB anomalies was also found to have a high prevalence of OSD and tethered cord[7]. In the present study, patients with ARM were screened for KUB abnormalities, and these were done in 185 patients. Among these, 65 (31%) urinary tract abnormalities were found in patients with ARM (n=204). The fourth most common KUB abnormalities comprised 44 (23.8%) hydronephrosis, 26 (14.1%) vesicoureteric reflux, 18 (9.7%) hydroureter, and 14 (7.6%) renal agenesis. Patients with ARM associated with KUB anomalies had a statistically significant correlation with OSD according to the Pearson chi-square test (p=0.001).

The methods utilized for radiologic screening for spinal dysraphism, such as a plain lumbosacral X-ray, U/S, and spinal MRI, were compared. Each radiologic modality was found to have some limitations. Plain lumbosacral X-ray could demonstrate sacral hypo-development, spina bifida, abnormal spinal curve, presacral mass, and hemivertebrae, but incomplete ossification in a plain lumbosacral X-ray in a neonatal spine would be difficult to ascertain. Plain lumbosacral X-rays that revealed sacral hypo-development, presacral mass, or sacral hemivertebrae had a higher prevalence for indicating OSD and tethered cord[7]. In the present study, vertebral spine anomalies were detected by a plain X-ray of the spine in only 29 patients (19%). Plain spinal X-rays could

detect some spinal anomalies with a sensitivity of 51.7% (95% CI 34.4 to 68.6), specificity of 88.4% (95% CI 81.5 to 93), accuracy of 81.3% (95% CI 74.3 to 86.3), and positive predictive value of 51.7%. Thus, a plain spinal X-ray was not a good modality for screening spinal anomalies in patients with ARM and would not be able to identify spinal cord lesions.

High-resolution ultrasonography is a rapid, safe, non-invasive method of evaluation because of the associated acoustic window provided by the spina bifida and incomplete ossification of the posterior elements in infants. Moreover, it can be done at the bedside and does not require sedation and in the authors' hospital. Thus, it was already being used to screen for genitourinary anomalies. The limitations of the U/S technique are operator dependence and the inability to identify a tethered cord in the newborn period. The limitation of U/S is the patient age for examination. Ultrasonography is highly accurate and effective in neonates when the spine is still cartilaginous. In a child who has ossified vertebra, MRI could more accurately identify abnormalities of the spinal cord. U/S can identify either spinal cord anomalies or spine anomalies. In the present series, among 65 patients that underwent U/S, abnormal ultrasonographic findings were found in 16 patients (24.6%). The abnormal findings detected by ultrasonography (n=16/65) comprised nine (13.8%) tethered cord, seven (10.8%) syringomyelia, three (4.6%) filar cyst, and one (1.5%) each of thick filum terminale without tethered cord, spinal lipoma, spina bifida, or sacral hemivertebrae.

The advantages of spinal MRI are the lack of ionizing radiation and superb demonstration of the anatomy and pathology, but the disadvantages include the high cost of MRI, the need for sedation or general anesthesia to obtain good quality images, and difficulties in imaging in severe scoliosis. Some studies recommend that spinal MRI should be conducted in every case of ARM[1,4,6,9,10].

MRI can identify either spinal cord anomalies or spinal anomalies. Among 39 patients underwent spinal MRI in the present study, abnormal MRI findings were found in 27 patients (69.2%). Some patients had two or more spinal cord and spine anomalies. The pathologic spinal cord findings detected by MRI (n=27/39) were 16 (41%) tethered spinal cord, 12 (30.8%) spinal lipoma, 10 (25.6%) syringomyelia, two (5.1%) lipomeningomyelocele, two (5.1%) filar cyst/ perineural cyst, one (2.6%) lipomeningocele, and one (2.6%) a thickened fatty filum with a mild low-lying cord. MRI could detect other spine abnormalities as well. MRI was the best investigation and had a high sensitivity and specificity.

Tethered cord syndrome has neurologic deficits involving the lower extremities and anal and urinary sphincters. In the present study, most of the children with OSD were asymptomatic. The symptomatic patients presented with motor symptoms, scoliosis, orthopedic gait abnormalities, urinary incontinence, and fecal incontinence. The indication of operation for OSD was controversial. In the present study, 33 (16.2%) of 204 patients with ARM had a radiologic diagnosis of spinal dysraphism. Only nine patients (27.3%) had symptoms of neuromotor deterioration or urinary or fecal incontinence. Spinal cord operations were conducted in 17 patients (17/33=51.1%). Among nine symptomatic neuromotor deterioration or urinary or fecal incontinence, eight patients were operated on, except one with a symptom of fecal incontinence. Prophylactic operations were conducted in nine patients. The options of operative treatments depended on the spinal abnormalities and preoperative clinical symptoms. Most of the operations were for untethering the spinal cord (82.4%) and the excision of intradural lipoma (29.4%). Six of the eight symptomatic patients who underwent an operation showed improvements of their symptoms postoperatively, whereas two patients did not show improvements in urinary and fecal incontinence postoperatively. The results of the operations in the present series were in accord with Tuuha et al's study[11], who reported that neuro or motor functions were clearly improved following surgery in symptomatic patients, but bowel and urinary functions remained unchanged following the surgery. In the asymptomatic group operated for prophylactic reasons, no patient had a deterioration of their neurological signs and symptoms postoperatively. Of the 17 patients underwent surgical treatments, five patients (29.4%) had complications. One patient with CSF leakage developed pseudomeningocele and so the repair of pseudomeningocele was further performed. The other complications improved without the need for a second operation.

In the present study, screening the spinal cord of a patient before 24 months of age could allow detection in patients before they developed neuromotor deterioration or urinary or fecal incontinence (7.9%), which was much better than screening the spinal cord in patients after 24 months of age (37.5%), with statistical significance at $p<0.05$. Therefore, patients with ARMs should be screened for OSD before 24

months of age before neurological deterioration has occurred.

However, the present study had many limitations to note due to retrospective review and selection bias of radiologic investigations in each patient. The methods used in radiologic screening for spinal dysraphism, such as plain lumbosacral X-ray, U/S, and spinal MRI, could not be compared precisely.

Conclusion

A high prevalence of OSD was noted in patients with ARM. All patients with ARM should be screened for OSD. U/S in neonates is the first-line screening method. Spinal MRI is strongly recommended in cloaca malformation, rectobladder neck fistula, rectoprostatic urethral fistula, vestibular fistula, and in all patients with ARMs associated with KUB anomalies. The other types of anorectal malformation should also be screened by spinal MRI for OSD, if feasible.

Screening the spinal cord in patients before 24 months of age could detect OSD before neurological symptoms have developed. Therefore, patients with ARMs should be screened for OSD before 24 months of age. Neuro or motor functions were found to be clearly improved in symptomatic patients, but bowel and urinary functions remained unchanged following spinal surgery.

What is already known on this topic?

ARMs were definitely associated with OSD, which comprises tethered cord, diastematomyelia, neuroenteric cyst, and spinal lipoma.

If OSD is left unattended, deterioration involving neurologic deficits would progress. Prophylactic surgical untethering in patients with a tethered cord who had no neurological deficit would be of great benefit.

The rational for performing screening tests is still considered controversial.

What this study adds?

In the present series, the prevalence of OSD in patients with ARM was 16.2%. All patients with ARM should be screened for OSD.

The different types of ARM had different prevalence of spinal dysraphism. The more complex the ARM, the greater the prevalence of spinal dysraphism, especially for vestibular fistula, which had a high prevalence of spinal dysraphism (20.5%).

Patients with ARM associated with KUB anomalies had a statistically significant correlation with OSD.

A plain spinal X-ray was not a good modality for screening spinal anomalies in patients with ARM and would not be able to identify spinal cord lesions.

Spinal U/S in neonates is the first-line screening method.

Spinal MRI is strongly recommended in cloaca malformation, rectobladder neck fistula, rectoprostatic urethral fistula, vestibular fistula, and in all patients with ARMs associated with KUB anomalies. The other types of anorectal malformation should also be screened by spinal MRI for OSD, if feasible.

Neuro or motor functions were found to be clearly improved in symptomatic patients, but bowel and urinary functions remained unchanged following spinal surgery.

Screening the spinal cord in patients before 24 months of age could detect OSD before neurological symptoms have developed.

However, this study had many limitations due to retrospective review and selection bias of radiologic investigations in each patient. The methods used in radiologic screening for spinal dysraphism, such as plain lumbosacral X-ray, spinal U/S, and spinal MRI, could not be compared precisely.

Conflicts of interest

The authors declare no conflict of interest.

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