

Ultrasound of the Siren Syndrome (Sirenomelia) in the District of Bamako Apropos of a Case

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Abstract

Sirenomelia is a rare fetal malformation with more or less complete fusion of both lower limbs consistently associated with bilateral renal abnormalities. The positive diagnosis can be made antenatally by ultrasound during the first trimester of pregnancy but often before the 22 weeks of amenorrhea. The objective was to think about the possible diagnosis by ultrasound in front of the urinary anomalies and the oligoamnios during the morphological study of the second trimester of pregnancy. We report a case of sirenomelia in a 25-year-old lady as part of the routine prenatal assessment suspected and confirmed during the morphological ultrasound of the second trimester of pregnancy and confirmed after the termination of pregnancy in Mali in the Health Center of Reference of commune III of the district of Bamako. The morphological ultrasound made it possible to diagnose the malformation of the urinary tract, the amniotic fluid and to make the therapeutic decision of medical termination of pregnancy at the end of confirming the siren syndrome.

Keywords

Sirenomelia, Ultrasound, Fetal Malformation, Mali

1. Introduction

Sirenomelia is a fetal malformation characterized by a variable degree of fusion of the lower limbs. It is a rare form of caudal dysgenesis described for the first time by Rocheus in 1542 and then by Polfyr in 1553. Its prevalence is estimated at 1 per 100,000 live births. This pathology is not compatible with life and is much more often associated with other fetal malformations, especially renal [1] [2]. The positive diagnosis is therefore based on ultrasound, which visualizes the malformation and specifies the different associations of the sequence and it is in the first trimester (12 - 13 SA) that the diagnosis should be made, with the help of a probe. Endovaginal [3] very few cases have been reported in Africa, especially in Mali, with a single case in the Sikasso region [4] [5] [6]. We report our case of sirenomelia observed in Mali in commune III of the District of Bamako.

2. Observation

This was a 25-year-old woman with no medical-obstetrical or surgical history and no notion of loss of amniotic fluid or taking medication. No particular exposure. She had consulted for the morphological ultrasound as part of the routine prenatal check-up in the referral health center of commune III in the district of Bamako. Performed by a GE (General Electric) Logic7 type ultrasound scanner with color Doppler equipped with a multifrequency sector probe. The different sectional planes show a pregnant uterus containing a fetus with unsatisfactory mobility. The placenta was well inserted posteriorly with a normal thickness of 29.4 mm. Amniotic fluid is scarce. The cord was well visualized with two arteries and one vein. Cardiac activity was assessed with 154 beats per minute. The umbilical Doppler gives a normal piglet index of 0.74. The uterine Doppler gives a normal piglet index of 0.59 on the right and 0.56 on the left (Figure 1).

Biometry gave a gestational age of 23 WA + 1 day (Figure 2).

On the morphological level: the study of the cephalic pole shows the brain structures of normal symmetrical appearance with good sphericity of the skull of regular contours. The septum pallidum saw the lateral ventricles not dilated. The cerebellum measured at 27.2 mm. The view with 2 nostrils and the upper lip continues. The clean bone of the measured nose has 8.8 mm.

The spine is visualized over its entire length.

At the chest level: the 4 cavities were symmetrical, the septum intact, the departure of the large vessels was normal, the aorta at 5.4 mm, the pulmonary artery at 5.6 mm. The diaphragm was present. N terms of the abdomen: the abdominal wall was continuous, the liver, the gallbladder, and the normal stomach. The bladder is not seen, supposedly empty or absent. There was an agenesis of a kidney and the other kidney carrier of multiple cysts (Figure 3).

The segments of the upper and lower limbs have been poorly studied due to the severe oligo-amnios and the posture of the fetus.

The therapeutic decision was the interruption of pregnancy before the severe

oligo-amnios and the renal malformation of the agenesia and poly cystic kidney type associated with an absence of visualization of the bladder which is incompatible with life.

After the interruption of pregnancy, delivery by the low way, we observed: a fusion of the lower limbs associated with a shortening of the humerus and an anomaly of form and number of the fingers and toes making evoke a sirénomelie (**Figure 4**).

The mother is doing well physically. She is followed by a psychologist from the health center on the mental level for better socio-cultural integration.

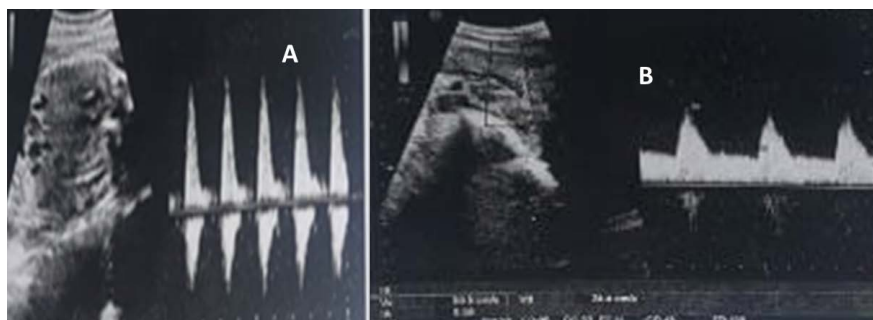


Figure 1. (A) and (B) show heart rate and uterine artery.

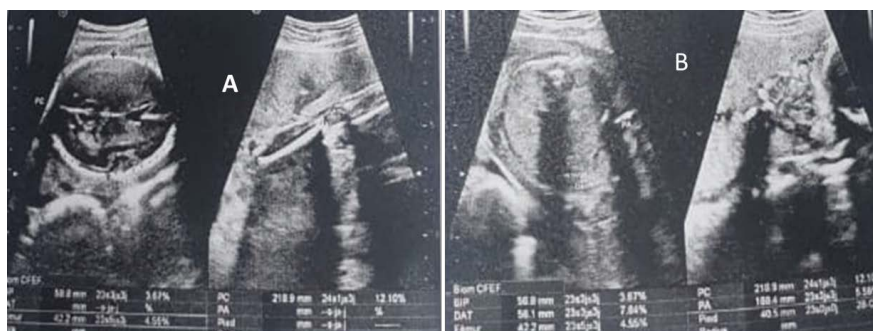


Figure 2. (A) and (B) show fetal biometrics.



Figure 3. Red arrow shows poly cystic kidney.



Figure 4. Photo of the fetus after termination of pregnancy.

3. Discussion

Siren syndrome or sirenomelia (Mermaid Syndrome), being a rare congenital pathology, usually presents associated anomalies, as in our case: a right bladder and renal agenes associated with a left renal polycystic. On the other hand, anal imperforation, absence of external or ambiguous genitals or even potter facies with anomalies of the vertebrae were not present in our case. Severe oligoamnios were present in our case as described in the literature [3] [7] [8].

More rarely, there are cardiovascular abnormalities, abdominal wall defects, pulmonary hypoplasia and thoracic malformations, and central nervous system abnormalities (hydrocephalus, anencephaly, craniorachischisis, etc.) [9] [10]. The association with a long bone anomaly such as the shortening of the humerus has been seen in our case; radial agenesis is rarely described in the literature [3] [11] [12]. Sirenomelia, apart from its prenatal diagnosis, is a malformation incompatible with life and survival after birth is very short. The positive diagnosis is therefore based on ultrasound, which visualizes the malformation and specifies the different associations. It is in the first trimester (12 - 13 SA) that the diagnosis should be made, with the help of an endovaginal probe while the amniotic fluid is still normally present, favoring the visualization of MI and its movements [2] [13] [14]. In case of doubt, an ultrasound around 17 - 18 SA is necessary, but usually, it is before the first warning sign that represents the oligo-anamnios (consequence of renal agenesis [13] [15]) that the diagnosis is made in the 2nd trimester. In our observation, he had not had a first trimester ultrasound like many pregnant women across Mali, despite the incredible accessibility of technical and financial means these days. This may most likely be due to the lack of information or the quality of the information provided to our pregnant women.

The examination looks for this unilateral or bilateral renal agenesis, the bladder, as well as the other malformations listed above. Amnioinfusion may be necessary to recognize fused lower limbs with little or no mobility. Color Doppler ultrasound is essential as it can circumvent the difficulty due to oligohydramnios [3] [16]. It studies the abdomino-pelvic vascularization, the aspect of the abdominal aorta (complete or partial atresia) and can make it possible to verify the absence of renal vessels. In renal agenesis alone, the aortic bifurcation is always observed [16] [17].

On ultrasound, sirenomelia can cause diagnostic difficulty with caudal regression syndrome (CRS). The distinction between these entities is important, especially for genetic counseling and the obstetrical future of these patients [3]. On ultrasound, in the CRS, we will find the classic V-shaped aspect of the femurs in abduction, the fetus in Buddha, normal abdominal fluid, no renal abnormality, and the presence of the two umbilical arteries. Maternal diabetes should be sought in principle [3]. On the other hand, a few cases of diabetic mothers have been described in sirenomelia in the literature, 10% to 15% of cases [1]. Mermaid syndrome is a sporadic malformation (unlike caudal regression syndrome [3] [18]).

4. Conclusion

Sirenomelia is a rare congenital malformation that must be considered as much as possible on morphological ultrasound in the face of an anomaly of the urinary tract and a significant drop in the quantity of amniotic fluid preventing total fetal exploration. A therapeutic decision of medical termination of pregnancy in the face of these renal and bladder abnormalities incompatible with life confirmed the strong suspicion of fetal malformation of the type (mermaid syndrome).

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

Consent

The parents were informed and they gave their informed consent.

References

- [1] Lubala, T.K., Mukuku, O. and Mutombo, A.M. (2014) Sirénomélie (Mermaid Syndrome): Description du premier cas Congolais et revue de la littérature. *Pan African Medical Journal*, **17**, Article 162. <https://doi.org/10.11604/pamj.2014.17.162.3934>
- [2] Valenzano, M., Paoletti, R., Rossi, A., Farinini, D., Garlaschi, G. and Fulcheri, E. (1999) Sirenomelia, Pathological Features, Antenatal Ultrasonographic Clues, and a Review of Current Embryogenic Theories. *Human Reproduction Update*, **5**, 82-86. <https://doi.org/10.1093/humupd/5.1.82>
- [3] Ladure, H., D'Hervé, D., Loget, P. and Poulain, P. (2006) Diagnostic anténatal d'une

- sirénomélie. *Journal de Gynécologie Obstétrique et Biologie de la Reproduction*, **35**, 181-185. [https://doi.org/10.1016/S0368-2315\(06\)76393-1](https://doi.org/10.1016/S0368-2315(06)76393-1)
- [4] Ugwu, R.O., Eneh, A.U. and Wonodi, W. (2011) Sirenomelia in a Nigerian Triplet: A Case Report. *Journal of Medical Case Reports*, **5**, Article No. 426. <https://doi.org/10.1186/1752-1947-5-426>
- [5] Morfaw, F.L. (2012) Sirenomelia in a Cameroonian Woman: A Case Report and Review of the Literature. *Journal of Clinical Case Reports*, **2**, Article 183. <https://doi.org/10.4172/2165-7920.1000183>
- [6] Cissouma, A., et al. (2021) Sirénomélie (Mermaid Syndrome): Description du premier cas dans la région de Sikasso, Mali et revue de la littérature. *Journal of African Clinical Cases and Reviews*, **5**, 439-442.
- [7] Ocelli, B., Vaast, P., Subtil, D., Metzger, N., Valat, A.S., Devisme, L. and Puechm, F. (1998) La sirénomélie: Revue de la littérature à propos de trois cas. *Médecine Foetale et Échographie en Gynécologie*, **34**, 24-31.
- [8] Nunez, M.J., Arcienega, E.G. and Bustillos, A.J. (2003) Sirenomelia. *Revista del Instituto Médico Sucre*, **68**, 67-70.
- [9] Plendl, P.J. (2003) Die Symmelie (sirenomelie) bei Mensch und Tier: ein komplexes Fehlbildungs-syndrom; dargestellt an vier neuen Fällen unter Berücksichtigung der gesamten bisher bekannten Literatur. Justus-Liebig-Universität Gießen, Gießen. <http://geb.uni-giessen.de/geb/volltexte/2003/1037/>
- [10] Halder, A., Pahi, J., Chaddha, V. and Agarwal, S.S. (2001) Sirenomelia Sequence Associated with Craniorachischisis Totalis, Limb Reduction and Primitive Heart. *Indian Pediatrics*, **38**, 1041-1045.
- [11] Kulkarni, M.L., Abdul Manaf, K.M., Prasannakumar, D.G. and Kulkarni, P.M. (2004) Sirenomelia with Radial Dysplasia. *The Indian Journal of Pediatrics*, **71**, 447-449. <https://doi.org/10.1007/BF02725639>
- [12] Ferru, J., van Nerom, P.Y., Ferraz, F.G., Houcke, M., Delecour, M., Farriaux, J.P., et al. (1982) Ectrourie: À propos d'un cas. *Archives Françaises de Pédiatrie*, **39**, 703-706.
- [13] Carbillon, L., Seince, N., Largilliere, C., Bucourt, M. and Uzan, M. (2001) First-Trimester Diagnosis of Sirenomelia. *Fetal Diagnosis and Therapy*, **16**, 284-288. <https://doi.org/10.1159/000053929>
- [14] Monteagudo, A., Mayberry, P., Rebarber, A., Paidas, M. and Timor-Tritsch, I.E. (2002) Sirenomelia Sequence: First-Trimester Diagnosis with Both Two- and Three-Dimensional Sonography. *Journal of Ultrasound in Medicine*, **21**, 915-920. <https://doi.org/10.7863/jum.2002.21.8.915>
- [15] Sirtori, M., Ghidini, A., Romero, R. and Hobbins, J.C. (1989) Prenatal Diagnosis of Sirenomelia. *Journal of Ultrasound in Medicine*, **8**, 83-88. <https://doi.org/10.7863/jum.1989.8.2.83>
- [16] Sepulveda, W., Corral, E., Sanchez, J., Carstens, E. and Schnapp, C. (1998) Sirenomelia Sequence versus Renal Agenesis: Prenatal Differentiation with Power Doppler Ultrasound. *Ultrasound in Obstetrics & Gynecology*, **11**, 445-449. <https://doi.org/10.1046/j.1469-0705.1998.11060445.x>
- [17] Patel, S. and Suchet, I. (2004) The Role of Color and Power Doppler Ultra-Sound in the Prenatal Diagnosis of Sirenomelia. *Ultrasound in Obstetrics & Gynecology*, **24**, 684-691. <https://doi.org/10.1002/uog.1758>
- [18] Harika, G., Gabriel, R., Bory, J.P., Gaillard, D., Quéreux, C. and Wahl, P. (1995) La sirénomélie: Revue de la nosologie à propos d'un cas. *Journal de Gynécologie Obstétrique et Biologie de la Reproduction*, **24**, 49-51.