The disorders of sex development and the problems of their management in Togo

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Sir,

The disorders of sex development (DSD) are attributed to abnormal budding of genital height abnormal located urethra orifice and or at least a non-concrete gonad.[1] It constitutes a medico surgical emergency in children.

The DSD are always distressing and depressing situations for the patients and parents as well. An early diagnosis, a hormonal deficiency treatment and a genitoplasty diminish individual and familial psychological consequences of this malformation.[2] From 12 cases of DSD, we want to determine the frequency of the DSD, to list the different DSD and to identify the problems related to them in Togo.

On 6 years from January 2006 to December 2011, we managed 12 patients with DSD in Sylvanus Olympio Teaching Hospital of Lomé. The records of patients seen in consultation or operated provided the basis to meet the parameters studied include age, sex of rearing, the results of karyotype, antenatal diagnosis, type of DSD and any treatment received by the patient.

The annual frequency was two cases. The average age of the patients was 7.41 years (ranges: 8 days-18 years).

The Table 1 sums up the epidemiological, diagnostic and therapeutical characteristics of the patients.

This study reveals the various problems of DSD in Togo. The low-rate would be related to a lack of information concerning this malformation. In fact, we received a great number of intersexual patients following a consultation and free treatment campaign. The malformations in general are considered to be devilish or witchcraft manifestation in African population, children then rarely access to consultations.[3]

Various type of DSD are identified: the feminine pseudo hermaphroditism come first with 5 cases over 12. In general, the children are changed to girls.[4] The masculine pseudo hermaphroditism must be suspected when one or more of these elements are present: Hypospadias, a unit or bilateral default of testicular migration.[5] Those patients are raised in boys very often as it is in our study.

In the gonadic dysgenesis and Tuner syndrome, the raising sex is difficult to determine. Jaubert *et al.*[6] recommend to raise them in boys. However, it is preferable to raise them in girls because of the presence of vagina and uterus.[7]

The small number of doctors and the precarious tools in our country do not permit biologic analysis of the amniotic nor the karyotype witch is done at Cotonou (Benin) and

Table 1: Epidemiological, diagnostic and therapeutical characteristics of the patients

Patient No.	Age	Prenatal diagnosis	Sex	Genetic sex (Karyotype)	Type of DSD	Treatment
1	8 days	None	M	Not defined	FPH	Not yet operated
2	8 days	None	Not defined	Not defined	Not labeled	Not yet operated
3	13 days	None	F	F (46,XX)	FPH	Not yet operated
4	16 months	None	M	M (46,XY)	MPH	Urethroplastia Duplay
5	3 years	None	F	F (46,XX)	FPH	TUM+Vaginoplastia+Clitoridoplastia
6	4 years	None	F	M (46,XY)	MGD	Bilateral Castration+Feminizing genitoplastia
7	5 years	None	F	F (46,XX)	FPH	Feminizing genitoplastia
8	7 years	None	M	M (46,XY)	MPH	Urethroplastia (Duplay)
9	13 years	None	F	F (46,XX)	FPH	Vaginoplastia+Clitoridoplastia
10	16 years	None	F	46, X0	Tuner Syndrome	Not yet operated
11	17 years	None	M	F (46,XX)	FPH	Patient and parents refused a sex changing
12	18 years	None	F	F (46,XX)	FPH	Feminizing genitoplastia

DSD: Disorders of sex development, M: Male, F: Female, FPH: Feminine pseudo hermaphroditism, MPH: Masculine pseudo hermaphroditism, MGD: Mixed gonadic dysgenesis, TUM: Total urogenital mobilization

poor parents could not trip to Benin for the karyotype.

In developing countries, the diagnosis is rarely done at birth. The delay of diagnosis in our study was related to the ignorance of the pathologies by the midwives, the paediatric doctors and the limited financial means. However, in major cases in developed countries, DSD is mainly antenatal and worst neonatal.

The diagnosis errors on the sex that does correspond to the genetic sex are often source of psychosocial conflicts more drastic than the delayed diagnosis. Must the surgery strictly respect the raising sex in these situations? We think one must give priority to the sexual identity of the child. One must consider socio-cultural habits contextually. There were no psychosocial assistance and check-ups with the patients, because the multi-disciplinary crew is not organized for the treatment of some abnormalities. A born child with DSD is source of frustration, deception and anxiety for the parents. An ambiguous genitalia child develops personality and psycho-affective disorders when his sex is different from his genetic sex (two patients in our study). The same problems have been mentioned by Abbey *et al.* in USA.[8]

We conclude that the DSD set diagnostic, therapeutical, psychological problems as well as determination and declaration constraints. The limited number of specialised crew and tools, the ignorance of the DSD by parents and caretakers and the lack of financial means highly contribute to these problems.

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