AVIS 132

ETHICAL QUESTIONS RAISED BY THE SITUATION OF PEOPLE WITH DIFFERENCES OF SEX DEVELOPMENT

COMITÉ CONSULTATIF NATIONAL D'ÉTHIQUE
POUR LES SCIENCES DE LA VIE ET DE LA SANTÉ

AVIS 132

ETHICAL QUESTIONS RAISED BY THE SITUATION OF PEOPLE WITH DIFFERENCES OF SEX DEVELOPMENT

Opinion unanimously adopted by the members present on 19 September 2019

TABLE OF CONTENTS

SUMMARY

The Office of the United Nations High Commissioner for Human Rights considers that "*intersex people are born with sex characteristics (including genitals, gonads and chromosome patterns) that do not fit typical binary notions of male or female bodies.*" Following its reflection on the situation of such people concerned by differences of sex development (expression preferred by the CCNE to 'intersex people'), the CCNE in the present opinion, which was requested by the Ministry for Solidarity and Health, has principally focused its analysis on the care afforded to the children concerned, the support given to their parents, the evolution of medical practices, and how a consensus can be reached between the people concerned and physicians.

Manifestations of these heterogeneous and complex variations: the number of people affected varies greatly depending on the point of view—very many according to advocacy groups for intersex people (1.7% to 2%, or even higher according to some groups), whereas physicians view these variations as 'rare diseases,' which are seen in 0.02% of births. Most differences of sex development do not hinder assignment of sex and are not life-threatening.

Suffering and anger of the people concerned: the consultations highlighted the great suffering and anger of people who experienced early surgery and/or treatment. These people emphasize the physical, psychological, sexual, and social trauma experienced during childhood, adolescence, and adulthood, and their conflicts with many specialist physicians.

Unease of professionals: the consultations also revealed a feeling of unease among physicians, surgeons, and endocrinologists regarding accusations and questioning of their practices by advocacy groups. In this regard, the physicians emphasize that over the decades early surgical and/or hormonal interventions have declined.

Lack of evaluation of past interventions and scarcity of data hamper ethical reflection regarding the following realities: the irreversibility of interventions in young children in the absence of their consent; a situation hard to deal with for parents and children alike; fundamental differences of approach to the question. Nonetheless, the CCNE considers that ethical reflection should promote dialogue between all parties.

Complex ethical questions: in this type of situation, ethical reflection is subject to doubt, uncertainty, and embarrassment, notably concerning the determination of sex, a question that cannot simply be reduced to chromosomal, genetic, endocrine, and morphological data. Ethical reflection is also tested by what constitutes appropriate behavior of medical

teams, by tensions generated by differences of opinion between medical professionals and parents, by the place of consent and by the parents' understanding of the significance of consent and of intervention (or of non-intervention) for the child's development and choice. The CCNE considers that reflection prior to formulation of this opinion could help reduce these tensions, with a view to overcoming current disagreement, while reinforcing the support and information provided and developing training and research in this complex field.

Recommendations

1. There is a need to centralize consultations and interventions in a **single facility**, so as to facilitate the alignment of points of view and concentrate expertise in a limited number of people. The CCNE considers that it is fundamental that children and their parents are cared for at one of the four sites that compose the **National Reference Center for Rare Diseases-Genital Development (CRMR)**, by a specialized and experienced multidisciplinary team.

2. **The training** and **continuing education** of professionals, including in the psychological realm, should be improved, notably for those who work in obstetrics and neonatology departments. The CCNE suggests that the Ministry of Health, the French National Authority for Health, and the appropriate professional organizations should define good practices in this field.

3. **Medical and surgical procedures**, whether early or delayed, **should fulfill a medical need while offering a therapeutic benefit.** When a particular characteristic of the phenotype necessitates a delay in assignment of sex, unless there is a medical necessity the individual concerned, when mature enough to do so, should be involved in the choice of treatment offered if this treatment is irreversible and threatens the individual's physical integrity.

For all other differences of sex development for which a medical and/or surgical procedure is considered, the decision should be taken after consultation and deliberation by the multidisciplinary team of the reference center, with the consent of the parents and of the child, once that child is mature enough. The decision should be documented, included in the medical file, signed by all concerned, and inscribed in the register held under the responsibility of the Ministry of Health.

4. **Clear and understandable information** on the situation of people affected by differences of sex development should be provided. Information given by the reference center to parents and to the people concerned should take into account all the treatment possibilities and also non-treatment. Parents and those concerned should be given sufficient time to reflect before a decision is taken.

More generally, information should be included in school curricula (biology courses) and made publicly available.

5. **It is essential to set up exhaustive databases and to support international research in this field.** Complete documentation on all treatment measures, on non-treatment, and on potential consequences should be provided. The CCNE recommends the creation of a database of anonymous data in France and in Europe for research purposes, with the aim of defining national and international clinical and therapeutic directives.

6. **Support** should be given to children and their parents at the reference center, from birth to adulthood, as should open access to information, care, and advice.

7. To **promote essential dialogue** between health professionals, advocacy groups, the people concerned and their parents, the CCNE proposes the creation of a reference center and twenty centers of competence (which, in tandem with the reference center, can also provide the people concerned with follow-up close to their place of residence). The CCNE also proposes a **forum on practices and their consequences** to enable the different parties to discuss, exchange ideas, and express their points of view.

The CCNE also proposes organizing regular country-wide **interdisciplinary conferences** and **periodic meetings on ethical issues** open to all parties – health professionals, scientists, advocacy groups – so as to create conditions favorable to shared reflection on practices.

These initiatives, which are also open to civil society, should help **to advance society's perception of sex differences**.

INTRODUCTION

Long ignored, "intersex people"[1] have sought to emerge from their silence of the 2000s through the work done by their advocacy groups in the media to raise awareness of the difficulties they experience and of the sequelae of early surgical interventions. These advocacy groups made themselves known to the political world in the 2010s, at a time of increased legal reflection concerning notions of self-determination and consent that do not exclude litigation and claims for compensation.

There have been parallel developments in international thinking and in the stances adopted by national institutions[2], notably the Council of State in its study presented to the French Prime Minister in 2018, and a strong mobilization of advocacy groups that invited the CCNE[3] to study the ethical issues associated with differences of sex development[4] and their consequences.

Against this backdrop, which echoed recommendation No. 5 of the information report by the French Senate[5] that it should be consulted, the CCNE organized hearings between January and May 2019 from which observations and lines of thought emerged regarding care of the children concerned, support of their parents, changes in medical practices, and the modalities of forging a consensus between the people concerned and physicians. It was decided, at this stage, not to study questions linked to civil status, which was analyzed in detail in the Senate report (recommendations Nos. 14 and 15) and regarding which the CCNE considered its input would be quite limited.

The CCNE was then asked for an opinion by the Minister for Solidarity and Health, under the seal of the General Health Directorate (see *Appendix 1: request received on 1 July*

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[1] Rather than this term, the CCNE prefers "people with differences of sex development."

[2] Notably: Council of Europe (2013). Children's right to physical integrity. Report by M. Rupprecht, doc. 13297, 19 p.
Committee on the Elimination of Discrimination against Women (2016). Observations finales concernant the rapport of the France valant septième and huitième rapports périodiques. CEDAW/OHCHR, 20 p.
European Union Agency for Fundamental Rights (2015). The fundamental rights – situation of intersex people, 9 p.
Council of Europe (2017). Promoting the human rights of and eliminating discrimination against intersex people. Parliamentary Assembly, resolution 2191, adopted on 12 October 2017, 3 p.
Défenseur des Droits (2017). Le respect des droits des personnes intersexes. Avis No. 17-04, 17 p.
Conseil d'État (2018). Révision de la loi de bioéthique : quelles options pour demain ? Section of the report and studies; study adopted by the General Assembly of the Council of State on 28 June 2018, 262 p.

[3] The CCNE received a letter from a law practice asking it notably "*to emphasize the unwanted and intolerable nature of the harms suffered*" by intersex people, "*to provide suitable and effective responses to these*," "*to recognize intersex*," and "*to end all mutilating medical interventions on intersex people*."

[4] However, the CCNE synthesis report on the bioethics forum, published in July 2018, just notes: "*Respect for differences and personal singularities were also mentioned, in particular as regards homosexual and intersex people*," questions concerning differences of sex development having been little considered during the États généraux de la Bioéthique (Bioethics Forum), even though an advocacy group of intersex people was consulted on this occasion. Opinion No. 129 of September 2018 on the review of the bioethics laws does not address the subject.

[5] Senate (2017). Les variations du développement sexuel : lever un tabou, lutter contre la stigmatisation et les exclusions. Information report No. 441 by M. Blondin and C. Bouchoux, drawn up for delegation on the rights of women and equal opportunities between men and women, recorded on 23 February 2017, 236 pp.

2019), in the framework of reflection on the issuing of a decree designed to refer the children concerned systematically to the reference center for rare differences of sex development[6], to review the specifications of this center, and to define good practices.

The CCNE was asked three questions:

- *With regard to the principles of integrity, autonomy, and access to healthcare, excluding life-threatening situations, is it possible to differentiate procedures that can be practiced early and those that can be postponed, by taking into account diverse situations, the short-, medium-, and long-term aims of these procedures, and the absence of scientific certainty and of professional consensus? Should we consider that only those procedures that avoid a threat to life can be performed early, or that procedures that meet a medical necessity can justifiably be performed early, and if so for what purpose?*

- *In this context of scientific uncertainty, related to the scarcity of available studies and a lack of consensus, which ethical considerations should guide health professionals and parents in making a decision (therapeutic abstention or intervention) that guarantees their child's autonomy without constituting a lost opportunity?*

- *How should parents be given clear, full, and enlightening information, including on ethical considerations, and supported to guarantee that their decision is not dictated by the perception that their child has an anomaly that must at all costs be corrected and, what is more, quickly?*

To answer these questions, the present opinion is intended to formulate the observations and recommendations inspired by the CCNE's consultations and analysis of articles and studies on the subject.

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[6] See the decree of 9 May 2017 concerning the certification of reference centers for a rare disease or a group of rare diseases. Ministry of Social Affairs and Health.

OBSERVATIONS

1 A subject difficult to pin down because of extreme diversity in differences of sex development

The highly heterogeneous manifestations of these differences are fully inventoried in the medical literature, the most recent data coming from a 2018 international consensus statement,[7] which mentions over twenty types of large differences (see *also Appendix 4: List of the reference center DEV-GEN Lille 2017)*.

Given the complex nature for non-specialists of the classification used in this consensus statement, the CCNE proposes, for the purposes of the present opinion, to group the principal differences of sex development differently, schematically and necessarily imperfectly, to facilitate understanding of both the most frequent and the most exceptional cases. This classification is in line with that referred to by the physicians consulted and broadly agrees with that adopted by the Council of State in its study on the revision of the bioethics laws, as well as that mentioned by the Senate report.

The classification covers:

- 46 XX individuals with atypical genital development, principally represented by girls with congenital adrenal hyperplasia, which is the simplest form of hyperandrogenism and manifests as clitoromegaly, but which can be life-threatening because of adrenal insufficiency with "salt loss," ie, approximately 25 cases a year, fully inventoried;
- 46 XY individuals with, notably, partial or complete androgen insensitivity syndrome (in the latter: 1/20 000 to 1/60 000 births) or gonadal dysgenesis (1/20 000 births); partial androgen insensitivity syndrome, which can raise questions of sex assignment in infants, is a rare genetic syndrome characterized by a more or less marked insensitivity of tissues to testosterone and is responsible for a spectrum of clinical expression ranging from more or less complete masculinization to isolated infertility in a 46 XY individual;
- patients with 45 X/46 XY mosaicism and anomalies of the external genital organs (2/100 000 births);
- very exceptional cases of 46 XX/46 XY individuals with male and female organs;
- people with highly varied and rare non-hormonal anomalies (1/60 000 to 1/270 000 births).

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[7] M. Cools, A. Nordenström, R. Robeva, J. Hall, P. Westerveld, C. Flück, B. Köhler, M. Berra, A. Springer, K. Schweizer, V. Pasterski (2018). Caring for individuals with a difference of sex development (DSD): a consensus statement. *Nature Reviews Endocrinology*, 14, 415-429.

To this simplified enumeration can be added hypospadias, which are very frequent (1/150 births), in which the opening of the urethra is not at the tip but on the underside of the penis, at a variable distance from the tip.

Severe posterior hypospadias is often associated with other genetic, gonadal, or endocrine anomalies and are classified as rare (7/10 000). Less severe cases of hypospadia do not raise questions of sex assignment.

According to medical sources, very few people are affected by differences of sex development that complicate sex assignment, although there are no official precise data.

In very rare life-threatening cases physicians must intervene rapidly by means of hormonal treatment for certain forms of congenital adrenal hyperplasia or surgically for very serious physical malformations.

So, in **most cases of differences of sex development there is no problem of sex assignment.**

Whereas groups of intersex people prefer to avoid medical categorizations, which they believe "pathologize" intersexuality, it nonetheless seems difficult, in the context of the present opinion, to disregard these categorizations, since not all differences of sex development by any means are associated with a problem of sex assignment.

2 The terminology reveals the difficulty of reaching a unanimous understanding

What does the notion of intersex people[8] encompass and what do we understand by differences of sex development?

The United Nations Human Rights Office of the High Commissioner considers that: "*Intersex people are born with sex characteristics (including genitals, gonads and chromosome patterns) that do not fit typical binary notions of male or female bodies*"[9].

Considering oneself as an intersex person refers to situations difficult to identify because they are imprecise. The size of this group of people varies greatly depending on the point of view, as we lack clear references. It is very large according to groups of intersex people: 1.7 to 2% of births, even 10% of the population according to some groups[10]. The term intersex people used by militant groups stresses the physical characteristics and

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[8] See T. Lundberg, P. Hegarty, K. Roen (2018). Making sense of “intersex” and “DSD”: how laypeople understand and use terminology, 9, 161-173.

[9] https://www.unfe.org/wp-content/uploads/2017/05/UNFE-Intersex.pdf

[10] « *La question est avant tout celle d’une autodéfinition. L’intersexualité n’est donc pas un questionnement sur le genre binaire, c’est la conscience de ne pas faire partie de celui-ci* ». In V. Guillot (2008). Intersexes : ne pas avoir le droit de dire ce que l’on ne nous a pas dit qui nous étions. *Nouvelles questions féministes*, Éditions Antipodes, 27, 37-48.

excludes all notion of disorder or disease: according to these groups, intersex people do not pose a problem of medical management and are characterized by atypical sexual development, without necessarily uncertainty regarding the assignment of sex. Guidelines, protocols, and medical classifications such as the World Health Organization's International Classification of Diseases (ICD) and the Classification Commune des Actes Médicaux (CCAM) of the Caisse Nationale d'Assurance Maladie des Travailleurs Salariés explicitly advocate the "depathologization" of differences of sex development.

However, at this stage there is no willingness to remove the mention of sex from civil status. Advocacy groups are at one with physicians in distinguishing the theme of intersexuality from that of gender transitions in the context of transidentity, which refers to the choice of a sex by people who do not necessarily present differences of sex development[11].

In contrast, most French physicians show very limited acceptance of the notion of intersex and the medical profession refers to scientific categories, deeming that the question is one of "rare diseases" (0.02% of births) and applying the term intersex only to people whose sex cannot be assigned at birth, without performing additional tests, given that the presence of both female and male genital organs makes sex assignment difficult.

As a subject closely linked to sexual identity and sexuality, the semantics reveal that the terms used have subjective implications extending beyond description of the genital apparatus. The CCNE has chosen to employ the expression "**differences of sex development**," adopted in Switzerland[12], which does not involve the future sexual identity and excludes the notion of disease, but implies the existence of atypical sexual development.

3 The question of differences of sex development generates great tension

The CCNE's consultations shed light on the very **great suffering and anger of people who experienced early interventions**. These people emphasized the physical, psychological, sexual, and social trauma experienced in their childhood, adolescence, and adulthood, their conflicts with physicians specializing in these issues, and the diversity of situations.

Groups of intersex people refuse the "medicalization" of their situation, virulently criticize hormone treatments and surgical interventions implemented without the consent of the person concerned, and demand an end to all non-life-saving interventions without the

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[11] However, this distinction does not exclude a convergence in the expression of claims by the LGBT and intersex movements.

[12] Swiss National Advisory Commission on Biomedical Ethics (2012). On the management of differences of sex development. Ethical issues relating to "intersexuality". Opinion No. 20/2012, 28 p., Bern, https://www.nek-cne.admin.ch/inhalte/Themen/Stellungnahmen/en/NEK_Intersexualitaet_En.pdf.

consent of the children concerned[13]. The parents are mentioned little in their discourse, suggesting that the parents are implicitly excluded from any power of decision. Only the organization Surrénales, which comprises parents and people with congenital adrenal hyperplasia, voiced an opposing point of view, and also stressed that its members do not self-identify with the term intersex.

The consultations also shed light on the **difficulties experienced by physicians,** surgeons, and endocrinologists when faced with accusations by intersex groups and **questioning about their practices**. Their past actions relate to a given context defined by the recommendations then in force, which they recognize, but they stress the fact that over the decades their understanding of the subject has evolved and that early surgical and hormonal interventions have declined considerably[14]. However, in certain situations, the contribution of early interventions seems to them fundamental for the construction of the sexual identity of the children and/or for surgical reasons, whether or not there are difficulties in assignment of sex. Other French and foreign physicians consulted voiced opinions closer to those of intersex groups.

Everyone's testimony, whether painful, indignant, or defensive, nourished the reflection of the CCNE, which also relied upon the available documentation to identify the ethical issues in these situations and to make proposals focused on the search for a consensus. The foreign specialists consulted greatly enriched this reflection. **The lack of consensus regarding the solutions** to **adopt is undeniable.** The greatly differing points of view of the people concerned and of health professionals reveal both the sensitivity and complexity of the subject and render certainties impossible. The subject cannot be grasped in the same way when considering the points of view of groups of intersex people, physicians, parents, and the children concerned[15].

One constant emerges from the consultations: the **burden of the trauma experienced by the parents** at the birth of a child whose sex is difficult to determine with confidence because the child does not present all the signs of conformity with the expected male or female sexual characteristics. Uncertainty regarding the child's sexual identity creates a very difficult situation for the parents. Beyond these very unusual situations, the suffering of the parents is also immense when a child, without presenting a problem of sex

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[13] B. Moron-Puech (2017). Le droit des personnes intersexuées. Chantiers à venir. *La Revue des droits de l'homme*. 11, 29 p. DOI : 10.4000/revdh.2815.

[14] P.D.E. Mouriquand, D.-B. Gorduza, C.-L. Gay, H.F.L. Meyer-Bahlburg, L. Baker, L.S. Baskin, C. Bouvattier, L. Braga, A.C. Caldamone, L. Duranteau, A. El Ghoneimi, T. W. Hensle, P. Hoebeke, M. Kaefer, N. Kalfa, T.F. Kolon, G. Manzoni, P.-Y. Mure, A. Nordenskjöld, J.L. Pippi Salle, D.P. Poppas, P.G. Ransley, R.C. Rink, R. Rodrigo, L. Sann, J. Schober, H. Sibai, A. Wisniewski, K.P. Wolffenbuttel, P. Lee (2016). Surgery in disorders of sex development (DSD) with a gender issue: if (why), when, and how? *Journal of Pediatric Urology*, 12, 139-149. DOI: 10.1016/j.jpurol.2016.04.001.

[15] M. Mottet-Caisson (2019). Les variations du développement sexuel. Problème public émergent aux enjeux complexes. Diplôme de Master en Sciences sociales de l'EHESS ; mention : recherches comparatives en anthropologie, histoire et sociologie, Marseille, 154 p.

assignment, has singular sexual characteristics. The unexpected intrusion of the difference distresses the parents who wonder about the future of their child in a societal context marked by the gender binary.

Several expressions of suffering are voiced in different timeframes: the parents' suffering, which starts at the birth of the child, the suffering of the children when they become adults which is expressed through their experience, and the suffering of physicians in their reflections upon medical ethics and the evolution of practices.

Beyond this constant, how can these tensions be addressed from an ethical point of view? Reflection on this must deal with the following realities:
- the absence of sex assignment or ambiguity therein which is hard to live with for parents and children alike;
- the irreversibility of interventions on young children;
- the suffering and questioning of the parents and children;
- the impossibility of consent from a young child;
- the fundamental divergences in approaches to these questions.

These divergences span the medical world. Recommendations (care protocols or guidelines) are rare. There is no unanimity in France on the adoption of certain international recommendations that tend to exclude early interventions. These divergences complicate the relations between groups of intersex people and most French surgeons and endocrinologists: the former deny that surgical intervention and hormone treatment benefit the construction of the child's sexual identity and decry their long-term physical and mental sequelae, whereas physicians justify the interventions in terms of their corrective and functional purposes, and, in some cases, assert that for psychological reasons the body must, insofar as possible, correspond to the sexual identity.

The psychologists consulted referred to the crucial issue of sex assignment and of the absence of ambiguity of the sex organs for the establishment of parent-child relations, the quality of which is essential from birth. However, how can parental and societal perceptions evolve towards unconditional acceptance of these children if the only solution proposed is conformity, both rapid and radical, with the established norm?

Are endocrinology and surgery indispensable in aiding the subjective construction of sexual identity? How can an answer to this question be found in a context where tensions are high, when what is involved are irreversible interventions on the body of an infant or young child whose future judgment of such actions in adulthood cannot be foreseen? These tensions should, however, be eased by the development of **training, support, information, and dialogue.**

4 Lack of evaluation of past interventions and of data make ethical reflection difficult

A 2016 update[16] of the consensus statement of 2006[17], which revised previous practices of rather systematic interventions, inspired notably by John Money[18], a psychologist and theoretician of gender, highlighted the personalized approach and "*honest and open communication with patients and parents*." This update revealed that the "*Timing, choice of the individual and irreversibility of surgical procedures are sources of concerns. There is no evidence regarding the impact of surgically treated or non-treated DSDs [disorders of sex development] during childhood for the individual, the parents, society or the risk of stigmatization*."

The objective and highly varied criteria, such as the influence of society, religion, and culture, mental and physical health, fertility and sexuality, past and present care, and subjective criteria, like social integration, adaptation, autonomy, quality of life, satisfaction regarding medical care and notably insertion in society are poorly understood and are deserving of more numerous systematic, scientific, and multidisciplinary studies.

Surgeons, who continue to operate early on children with differences of sex development, assert that surgical techniques have advanced greatly and result in fewer sequelae, in particular scarring (changes to the techniques of surgery of the clitoris, vaginoplasty), and that they operate much less than in the past (great caution concerning gonadectomy, postponement of some interventions).

But the CCNE has no precise information on the current number of operations in the first years of life or their nature, other than for people with congenital adrenal hyperplasia. **This lack of accurate benchmarks** and of studies of how people who have been operated on perceive their quality of life **means that it is not possible ground ethical reflection in unequivocal data.**

5 The role of the parents is fundamental: ethical reflection should help consolidate this role

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[16] P.A. Lee, A. Nordenström, C.P. Houk, S. F. Ahmed, R. Auchus, A. Baratz, K. Baratz Dalke, L.-M. Liao, K. Lin-Su, L.H.J. Looijenga, T. Mazur, H.F.L. Meyer-Bahlburg, P. Mouriquand, C.A. Quigley, D.E. Sandberg, E. Vilain, S. Witchel and the Global DSD update Consortium (2016). Global disorders of sex development update since 2006: perceptions, approach and care. *Horm. Res. Paediatr*. DOI: 10.1159/000442975, 23 p.

[17] I.A. Hughes, C. Houk, S.F. Ahmed, P.A. Lee, LWPES/ESPE Consensus group (2006). Consensus statement on management of intersex disorders. *Journal of Pediatric Urology*, 2, 148-162. DOI: 10.1016/j.jpurol.2006.03.004.
P.L. Lee, C.P. Houk, S.F. Ahmed, I.A. Hughes (2006). Consensus statement on management of intersex disorders. *Pediatrics*, 118, e488-e500. DOI: 10.1542/peds.2006-0738.
Consortium on the management of disorders of sex development (2006). Clinical guidelines for the management of disorders of sex development in childhood. Intersex society of North America, 63 p.

[18] J. W. Money (1952). Hermaphroditism: an inquiry into the nature of a human paradox. Harvard University, Cambridge, Ma, USA. Ph.D., part 1 (231 p.) and part 2 (207 p.). See also: J. Colapinto (2000). As nature made him: the boy who was raised as a girl. HarperCollins Publishers, New York, USA, 289 p.

In the light of what the people concerned say, ethical reflection should help to develop the roles of the parents, advocacy groups, and health professionals, in view of the scientific uncertainty regarding the benefit of irreversible interventions[19]. Some previous practices inflicted on people with differences of sex development resulted in sequelae that were irreversible both physically and psychologically. The current ethical perspective leads to reflection on the basic rights of the individual, notably the right to respect of physical and mental integrity, and the rights to information and to self-determination.

Any decision is complicated by the difficulty of obtaining fully informed parental consent, which is extremely dependent on the information given, the diagnosis, and the possible prognoses, by the impossibility of obtaining the child's consent, and by the diversity of points of view.

The role of the parents is fundamental in the construction of the child's identity.
Parenthood is constructed throughout pregnancy and during the infant's early days, and the parents, whose role in the fate of newborns is essential, must make crucial decisions, regardless of whether there is a prospective intervention. Yet they often know little or nothing of differences of sex development at birth, sexual differentiation being a scientifically complex subject which is still little discussed. Parents need time to discover and to adapt emotionally to the birth of their child. The child's future may initially seem very bleak to parents; the child that they discover is alien to their ideals and to the "imaginary child" that they were expecting. They may be highly tempted to want to alter the child's physical appearance to comply more with binary, sexual, and gendered representations. According to this conception, not only are the child's interests at stake, the aim being to reduce the trauma stemming from the child's confrontation with intersexuality, but the objective also is to reassure the parents regarding their child's sex and reproductive life. Fertility is much reduced in nearly all forms of differences of sex development. The ethical questions are very difficult because of the nature of the organs concerned, their symbolism, and the parents' cultural representations.

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[19] See K. Gueniche, M. Jacquot, E. Thibaud, M. Polak (2008). L'identité sexuée en impasse…À propos de jeunes adultes au caryotype XY nées avec une anomalie du développement des organes génitaux et élevées en fille. *Neuropsychiatrie de l'enfance et de l'adolescence*, 56, 377-385.
A.-M. Rajon (2008). Ce que nous apprennent les parents d'enfants porteurs d'ambiguïté génitale. *Neuropsychiatrie de l'enfance et de l'adolescence*, 56, 370-376.
J.C. Streuli, E. Vayena, Y. Cavicchia, J. Huber (2013). Shaping parents: impact of contrasting professional counseling on parents' decision making for children with disorders of sex development. *J. Sex Med*, 10, 1953-1960
M. Gardner, D.E. Sandberg (2018). Navigating Surgical Decision Making in Disorders of Sex Development (DSD). *Front. Pediatr.*, 6:339. DOI: 10.3389/fped.2018.00339.

RECOMMENDATIONS

1 Given their rarity, cases should be managed in specialized facilities

The CCNE shares the intentions expressed in the letter of referral on the need to centralize consultations and interventions in a single facility, to expedite the alignment of points of view and to concentrate expertise in a limited number of people.

It seems **essential** that a specialized and experienced multidisciplinary team **should care for children and their parents at one of the four sites of the reference center** (CRMR) **for rare diseases relating to genital development**, so as to help the parents welcome their child and create the conditions needed to establish the parental bond[20]. Centralization of cases is essential, because of their very great specificity and the low number of critical cases. Know-how, a multidisciplinary approach, and a sufficient number of cases to foster discernment in decision making justify the one-by-one examination of all cases centralized at the reference center, which can also call upon the twenty centers of competence that have developed particular know-how in these fields. Care by staff with little or no training should be avoided at all costs. However, it is strongly recommended that the four sites further align their practices, in line with what is recommended hereafter, to reach a joint position that respects good practices, without excluding the questions inherent to all medical practice.

Through its four sites, the reference center should combine **specific skills in an interdisciplinary setting** that brings together physicians and experts in the disciplines concerned (gynecologist-obstetrician, sonographer, neonatologist, pediatric surgeon, endocrinologist, geneticist, child psychiatrist, psychologist, ethicist, clinical geneticist, urology surgeon, andrologist, legal expert, sociologist). Ethical reflection and deliberations on good practices are needed. Groups of intersex people should participate in the activities and in the definition of objectives. It is important to suggest to parents that they meet such groups and also families who have similar experiences, as they can offer appreciable help.

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[20] See notably: F. Phan-Hug, C. Kraus, A. Paolini-Giacobino, F. Fellmann, S-A Typaldou, F. Ansermet, L. Alamo, N. Eggert, O. Pelet, Y. Vial, V. Muehlethaler, J. Birraux, P. Ramseyer, S-C Renteria, A. Dwyer, N. Pitteloud, B. Meyrat (2016). Patients avec variation du développement sexuel : un exemple de prise en charge interdisciplinaire. *Rev. Med. Suisse*, 12, 1923-1929.
H.F.L. Meyer-Bahlburg (2008). Lignes de conduite pour le traitement des enfants ayant des troubles du développement du sexe. *Neuropsychiatrie de l'enfance et de l'adolescence*, 56, 339-344.

2 Training should be improved

At a time when medical teaching programs are under review, the various dimensions of differences of sex development should be included in the initial and continuing training of health professionals.

The quality of the announcement made to the parents that their child has a difference of sex development is essential, so it is imperative that sonographers and personnel in the maternity, neonatology, and pediatrics units be trained in providing such information in as pertinent a manner possible, before the families are referred to the reference center. All consultations with experts lead to this observation: the choice of words on such a sensitive subject is absolutely determinant for the life of the family from the child's birth and over the very long term.

Prenatally, the obstetrician and midwife are the first concerned: they should inform without advancing a diagnosis and refer the parents to those qualified to do so. Only a Multidisciplinary Prenatal Diagnostic Center can provide complete information on the discovery of a difference of sex development, at the time of prenatal tests or at birth. Neonatally, midwives, obstetricians, and pediatricians must refer families to the reference center. At birth, the medical teams must understand the care relationship while respecting good professional practices, pay special attention to the words they use, and help parents overcome their anxiety by putting them in touch with health professionals familiar with this type of situation. Given the complex and sensitive nature of the problems and the strict requirements that the multidisciplinary decision must satisfy, the CCNE suggests that the Ministry of Health, the Haute Autorité de Santé (French National Authority for Health), and other qualified professional organizations should establish good practices for the training and professional development of the specialists involved.

3 To take action or not?

The Minister's first two questions to the CCNE related to whether it is possible to differentiate between procedures that can be implemented early or postponed, taking into account the diversity of situations, the short-, medium-, and long-term aims of these procedures, and the lack of scientific certainty and of professional consensus. Can procedures implemented early because of a medical need be justified or should only those procedures that avoid a threat to life be justified?

A precise response from the CCNE is not possible, because the question encompasses technical considerations and medical approaches, notably empirical, that go beyond ethical considerations alone. The national protocol for the diagnosis and care (PNDS) of partial or complete androgen insensitivity syndrome posted on the Haute Autorité de Santé website[21] does not exclude all interventions, but exercises extreme caution: "*...however, it is not possible to consider all specific cases or complications, all therapeutic characteristics, and all hospital care protocols. No claim can be made for coverage of all possible care scenarios, nor can this protocol be a substitute for the individual physician's responsibility to the patient*."

How can we define the limits of interventions between what is medically necessary, what is medically preferable, and what is subject to questioning once we leave the medical world for the social sphere? At the heart of this question is respect of the individual's identity and difference. The uniqueness and specificity of each situation call for case-by-case analysis within a timeframe determined according to the child's best interests.

Legally, what the Council of State reiterates in its opinion on the revision of the bioethics law can be used as a benchmark for ethical reflection in this field. The aim is to balance different imperatives.

Article 3.1 of the United Nations Convention on the Rights of the Child emphasizes the best interests of the child: "*In all actions concerning children, whether undertaken by public or private social welfare institutions, courts of law, administrative authorities or legislative bodies, the best interests of the child shall be a primary consideration*."

Article 16-3 of the Civil Code derived from the law of 6 August 2004 concerning bioethics notes that: "*The integrity of the human body can only be violated in cases of medical necessity for the individual or exceptionally in the therapeutic interests of others*." The notion of consent is hard to grasp. The same article specifies that: "*the consent of the person concerned must be collected beforehand, except in cases where the medical condition requires a therapeutic intervention to which the person is not able to consent*." In principle, this text does not exclude very occasional medical decisions taken on a case-by-case basis in certain scenarios, even if the situation is not life-threatening.

Article 41 of the French Code of Ethics (article R. 4127-41 of the Public Health Code) asserts that: "*no mutilating intervention can be practiced without a very serious medical reason and, except for emergencies or impossibility, without informing the person concerned and without his or her consent*."

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[21] Haute Autorité de santé (2017). Insensibilités aux androgènes – PNDS Centre de référence du développement génital: du fœtus à l'adulte, 38 p. https://www.has-sante.fr/jcms/c_2818977/fr/insensibilites-aux-androgenes

Article 371-1 of the Civil Code assigns parental authority to the parents until the child's coming of age. While parents decide on treatments for their child according to article R. 1112-35 of the Public Health Code, article R. 4127-43 of the same code nonetheless underlines the predominant role of the physician, who "*is the defender of the health of the child whenever he or she deems that the interests of the child's health are poorly understood or protected by the child's entourage*." Article L. 1111-4 of the same code also takes into account the individual concerned by recommending that consent be systematically sought if the minor is able to express a wish and to take part in the decision making.

The procedures, whether implemented early or later, must meet a medical need while providing a therapeutic benefit. In cases where a particular characteristic of the phenotype necessitates a delay in the assignment of sex, unless there is a medical necessity the individual concerned, if of sufficient maturity, should be associated with the therapeutic choices offered whenever the irreversibility of the chosen treatments puts the individual's physical integrity at risk.

As a result, in the very particular context of scientific uncertainty regarding the advantages and drawbacks of early operations, the decision relating to these operations, when they are irreversible and remove or substantially alter a sex organ, should, in principle, only be taken by the people concerned, once they are able to make an informed choice. Whence the need to wait, except in emergencies or special situations, for the people concerned to be able to decide or, where appropriate, to be associated in an informed way with the decision making[22].

In these conditions, the self-determination of the child is essential and the child's physical integrity should be protected until he or she is of an age to decide, even though it is difficult to determine this age, which will vary from one person to another and as a function of the difference of sex development in question, because some irreversible interventions, if their physical and mental benefits have not been demonstrated, can be postponed until adulthood.

Given the scientific uncertainties, an indication motivated by psychosocial considerations alone currently does not justify an operation related to sex assignment practiced on the genital organs of a child incapable of discernment. A decision to operate on a sex organ must be the fruit of multidisciplinary reflection at one of the sites of the reference center. What is unknown is how the child, once an adult, will view the decision not to operate.

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[22]. B.M. Dickens (2018). Management of intersex newborns: legal and ethical developments. *Int. J. Gynecol. Obstet*., 143, 255-259. DOI: 10.1002/ijgo. 12573.
S.F. Ahmed, J.C. Achermann, W. Arlt, A. Balen, G. Conway, Z. Edwards, S. Elford, I.A. Hughes, L. Izatt, N. Krone, H. Miles, S. O'Toole, L. Perry, C. Sanders, M. Simmonds, A. Watt, D. Willis (2016). Society for Endocrinology UK guidance on the initial evaluation of an infant or an adolescent with a suspected disorder of sex development (Revised 2015). *Clinical Endocrinology*, 84, 771–788. DOI: 10.1111/cen.12857.

Since the 1990s, a certain number of very negative testimonies by American physicians[23] have led to criticism of the overly systematic nature of interventions.

For all other situations of differences of sex development that pose no problem of sex assignment, but for which the implementation of a medical and/or surgical procedure is under consideration, certain forms of congenital adrenal hyperplasia for example, it should be recalled, as did the Council of State, that an early surgical procedure must be a medical necessity for the individual concerned and provide a therapeutic benefit after study of the risk-benefit ratio. Such a medical necessity presupposes that compelling and firmly established reasons justify early surgical intervention before the child is able to participate in decision making, if the aim of the intervention is not strictly to avoid medical complications.

As a consequence, the final decision regarding an early surgical procedure should be taken by the multidisciplinary team of the reference center, after consulting the parents. The decision may be to postpone the intervention until the minor is able to participate in decision making, to express any suffering associated with the differences in sex development, and to measure the benefits and risks of the procedure envisaged [24].

The decision concerning the precise case referred to the reference center medical teams, whether for hormonal or surgical treatment, **should be recorded in a document, signed by the physicians and the parents and included in the medical file** and kept in an institution under the responsibility of Ministry of Health so that it can be made available at any time to the parents as well as to the individual concerned.

By adopting a case-by-case approach, which avoids "universalization" of the individual in a category, the **CCNE excludes any change to the civil code**, such as an addition stating that sex assignment procedures that are not urgent or needed to avoid a threat to life do not constitute a medical necessity.

The CCNE also excludes any change to the regulations concerning reimbursement for procedures performed on people presenting differences of sex development, once these

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[23] For example: M. Diamond, K. Sigmundson (1997). Sex reassignment at birth: long-term review and clinical implications *Arch. Pediatr. Adolesc. Med.* 151, 298-304.

[24] See J.A. Greenberg (2017). Legal, ethical, and human rights considerations for physicians treating children with atypical or ambiguous genitalia. *Seminars in Perinatology*, 41, 252-255.
C. Wiesemann, S. Ude-Koeller, G.H. Sinnecker, U. Thyen (2010). Ethical principles and recommendations for the medical management of differences of sex development (DSD)/intersex in children and adolescents. *Eur. J. Pediatr.*, 169, 671–679. DOI 10.1007/s00431-009-1086-x.

procedures are recorded in the Classification Commune des Actes Médicaux (article L. 162-1-7 of the social security code).

However, care protocols and good practice guides give physicians reference frameworks.

4 Which ethical considerations should guide health professionals in decision making?

The Minister's second question to the CCNE addressed the ethical considerations that should guide health professionals and parents in taking a decision (abstention or intervention) that guarantees the child's autonomy without constituting a lost opportunity.

The irreversibility of the intervention in a situation where the child's consent is lacking is, as we have said above, the main question. This is a sensitive issue for physicians because it concerns, on the one hand, a person who is not autonomous, that is an infant or young child, and, on the other hand, the parents, who are responsible for the child. Particularly sensitive is the question of the assignment of sex.

The difficulty is not specific to differences of sex development, since major surgery decided in agreement with the parents may have irreversible consequences: heart, gastrointestinal, or orthopedic surgery, correction of a cleft lip and cleft palate. But these situations usually involve a threat to life, which is generally not the case for interventions performed on children with differences of sex development.

This lack of necessity to avoid a threat to life modifies the relation between the painful and irreversible sequelae of such interventions and the resulting benefit. It is the risk-benefit ratio that is keenly discussed by those concerned, once they consider that the painful and definitive consequences, notably in terms of reproduction, outweigh the likely drawbacks of not intervening early. Interventions go well beyond medical treatment in the strict sense of the term since they penetrate to the very core of being a human. They can affect reproductive potential which, for some people, is a critical issue.

The evaluation of risks and advantages is fundamental and, in our current state of knowledge and bearing in mind the specificity of differences of sex development, should rule out any procedure that may constitute a lost opportunity. But this lost opportunity should have been assessed beforehand in light of the irreversible nature of an intervention, which can be measured, and, to a lesser extent, in light of other equally fundamental factors[25].

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[25] Such as the construction of an identity or a child's difficulty in positioning him- or herself in the classifications that prevail in society, but which are more difficult to evaluate and to some degree more changeable. Hence, a decision will weigh up factors that are measurable and others that are less easily measured.

In this type of situation, ethical reflection is subject to doubt, uncertainty, and embarrassment, notably regarding:

- questions concerning sex assignment; the question cannot always be reduced to chromosomal, genetic, endocrine, and morphological data;
- management within or between medical teams;
- tensions that may arise when the medical team and parents are of different opinions;
- parental understanding of the significance of consent;
- limits of delegating to the parents the choice of interventions and of sex assignment;
- outcome for the child and what could be the child's choice.

The dilemma is well illustrated by this quote[26]: "*Although clinicians have a duty to act in the best interests of their patients, objective standards of best interests for children may be hard to define in isolation. In DSD [disorders of sex development], there may be tensions between the potentially conflicting obligations of respecting a child or young person's right to make informed voluntary self-determined choices (autonomy) and providing treatment for the child that carries more benefit than harm. Moreover, any consideration of a child's best interests needs to include the obligation to respect the family and parent–child relationships*."

Note should be taken of the importance of the most logical and evident sex assignment, which in most cases is possible using the tests performed.

Should we recognize a right of a child or adolescent to make informed choices, in the name of the principle of autonomy, if he or she is imperfectly qualified to make decisions of this import, or should we instead renounce any intervention pending the person's coming of age?

The child's interests at each stage of life call for rethinking of the **parent-child relation, so that the** parents accept their children as they are and not as the parents wish they were, so that the parents know how to avoid the temptation of surgical interventions when they are unnecessary and how to build an untroubled relationship with their child.

In the interests of respect of ethical principles, emphasis should be placed on **how the parents are told**, as the information provided could influence parental positioning, and on the **support** of parents in such situations and their questions regarding **the development of their child's identity**.

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[26] C.E. Brain, S.M. Creighton, I. Mushtaq, P.A. Carmichael, A. Barnicoat, J.W. Honour, V. Larcher, J.C. Achermann (2010). Holistic management of DSD. *Best Pract. Res. Clin. Endocrinol. Metab.,* 24, 335–354.

Ethical reflection cannot overlook the need to rethink relations between the medical and paramedical personnel concerned, or the relations between them, researchers, advocacy groups, parents, and children. **The question of the relationship to society** is also essential. The people concerned live in a society that has a certain view of what is deemed atypical. A societal view which, currently, is unaccepting of differences of sex development and can be harmful and lead to forced normalization.

The CCNE agrees with the ethical guidelines of the Belmont Report[27] concerning the protection of human subjects of research: non-harm, beneficence, relevance (medical and therapeutic necessity), respect of private life and of autonomy, collection of free, informed consent.

5 Training and support are needed

The Minister's **third question** to the CCNE concerned how parents should be provided with clear, complete, and enlightening information, including on ethical considerations, and how they should be supported so as to guarantee that the decision is not dictated by the perception that their child has an anomaly which should, at all costs, be corrected.

The CCNE therefore identified **information and support** as two of the four basic pillars of care, as the parents' consent must be informed. In this regard, a fully documented article by Katrina Roen[28] highlights the balance between a medical approach that is purely physiological and an approach that is more psychosocial.

Consent can only be informed if the information given to the parents is fair, clear, and understandable, ie, accessible to their degree of comprehension. The way the decisions to be taken are presented, whether or not an intervention is being considered, is determinant in enabling parents to establish a relationship with their child and to accept that child as is. It is also fundamental for the provision of good care by physicians. Technical terms may further traumatize parents, whereas information that leaves room for psychology and sociology can help them see the problem in its entirety and participate in decision making or be more accepting of the decision. The emotional burden associated with the diagnosis is such that the information cannot be "black and white."

The information given by the reference center should encompass all possible short- and long-term physiological and psychological consequences of a treatment or of a lack of treatment, without neglecting scarring problems and scientific uncertainty regarding the benefits of interventions. The information should be guided by the principles governing

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[27] Office for Human Research Protections (1979). The Belmont Report. Ethical principles and guidelines for the protection of human subjects of research. U.S. Department of Health, Education and Welfare, 10 p.

[28] K. Roen (2019). Intersex or Diverse Sex development: Critical Review of Psychosocial Health Care Research and Indications for Practice. *The Journal of Sex Research*, 56, 511-528.

medical procedures that may be performed and procedures not performed in response to a medical necessity. The parents, and later the children when they reach an age where they understand or can be included in decision making, should be given enough time to reflect before a decision is taken; a rapid decision can only be envisioned in medical emergencies. The question of the declaration of civil status should not lead to hasty decisions. The information will be all the more credible when it is voiced by different members of the multidisciplinary team, thus enabling the parents to understand the question from the viewpoints of the various medical specialties concerned. The information should emanate from endocrinologists and surgeons as well as from psychologists and psychiatrists specialized in this type of question. As this now seems to be the case at the sites of the reference center, the parents will only benefit from being informed of the existence of advocacy groups of people with differences of sex development and from being encouraged to contact parents familiar with this type of experience.

Some parents may feel unable to live with a child who presents ambiguous sexual characteristics.

Here too the information cannot be one-way and the medical teams should indicate the risks, controversies, and absence of unanimity. Parental distress should not be overlooked: refusing all interventions will in certain cases impact the relationship between the parents and the child. External, even internal, malformations of the genital organs may create such confusion regarding sex assignment at birth that the parents will not tolerate non-intervention. The medical teams should try to explain that major and complex surgery intended to make the genital organs conform to the parents' wishes will not necessarily facilitate the construction of the identity of the child who, once an adult, may reproach them for having agreed to painful and irreversible operations.

In these situations, even though the surgeons do not plan to operate, the parents should be clearly told that such procedures done without the child's consent may have serious consequences. A risk-benefit ratio as accurate as possible should be established.

If an irreversible intervention is medically justified, the decision should be taken by the physicians responsible for the child's care, after consultation and reflection by the multidisciplinary team in a reference center, in agreement with the parents and the individual concerned, if the individual is old enough to voice an opinion, and after provision of information and discussion[29].

This information should not be occasional, but rather should be renewed well beyond the birth of the child, during regular consultations.

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[29] Notably, the right to revoke a decision at any time.

For medical care required by an individual throughout childhood and adolescence, the parents should be referred to the twenty centers of competence attached to the reference center, when it appears unnecessary that the follow-up be permanently conducted in this reference center.

Support should be provided at the reference center from birth to adulthood, and access to information, care, and advice should remain open. Children will need psychological and social support so that, on the one hand, they can speak of their situation and, on the other hand, they can take part in decision making concerning themselves, once they are capable of discernment. Specialized psychological follow-up should be offered to parents throughout their child's childhood and adolescence, so that they are not left to their own devices, do not depend solely on consultations with pediatric physicians, endocrinologists, or surgeons, and can talk about the problems they encounter. It is also advisable for the reference center to stay in touch with the children concerned once they reach adulthood, so as to continue to provide support if requested and to document the cases, notably by studying the questions that arise in the event of non-intervention.

The advice and support should not overlook the child's worries and needs, or those of the parents, which can vary greatly depending on the diagnosis, the prognosis, and the family situation. Scrupulous analysis of the situation therefore constitutes a necessary and indispensable pre-condition.

6 A database should be set up to help in decision making

Databases should be set up so as to follow advances in the field of differences of sex development and to undertake discussions based on accurate and reliable studies.

Complete documentation must be provided on all treatment measures (surgery and hormone therapy), non-treatment, and any consequences thereof. This documentation is made necessary by the possibility of side effects throughout the lives of people with differences of sex development, regardless of whether or not they were treated. These people should also retain the right of access to the treatments they received or to the decisions that led to an absence of treatment. The documentation must be kept for at least 40 years and should only be accessible to the people concerned. After anonymization, the data kept in this way will also be a precious source for researchers and physicians, enabling optimization of the medical and psychosocial approaches to each case of differences of sex development.

This is why the CCNE recommends the **creation of databases in France and in Europe for research purposes**, which will allow studies that help define therapeutic guidelines in France and internationally. These databases, whose confidentiality must naturally be subject to vigilant protection, could in time enable a reduction in the divergences of approach between the different centers specialized in the treatment of differences of sex development.

7 Promoting dialogue: creation of structures for dialogue would enable a move from disagreement to consensus

After these reflections and proposals, it appears necessary to return to what most clearly emerged from the consultations, namely, the disagreement between the people concerned and most physicians, and to a lesser degree the disagreement among physicians.

Creation of structures for dialogue is an ethical priority given **that perceptions must change regarding the question of differences of sex development**, in a society where many young people do not have the same assumptions as their elders regarding questions related to sex or gender and who are moving, admittedly not always unanimously, towards greater acceptance of differences, and even greater gender fluidity. Tolerance in this regard should be encouraged by information campaigns. It is without doubt necessary, regarding these situations, to move towards another vision of sex differences, one which also affects symbolic points of reference. But this desirable change is also linked to a new medical approach and should first and foremost affect those most concerned, many of whom cannot meet without clashing, sometimes violently.

In 2019, there is a lack of consensus on delimiting the scope of differences of sex development and on the vocabulary employed, a lack of dialogue between health professionals and groups of intersex people, and a lack of agreement on good practices in the interests of the child between medical teams and sometimes between the four sites of the reference center. In addition, other professionals with particular skills and expertise may voice different points of view, and militant groups and people who have undergone surgery have initiated court proceedings. The CCNE therefore proposes:

- to set up, with the reference center and its twenty centers of competence, a **forum on practices and their consequences**, to allow the people concerned, parents, health professionals, legal experts, and others involved in this field to exchange ideas, discuss, and hear different points of view;

- to organize regular national **interdisciplinary conferences and periodic conferences on ethical issues** for health professionals, patient groups, groups of intersex people, as well as specialists in ethics, the law, and gender studies, so as to create the conditions for joint reflections on practices. These meetings could enable major advances and encourage changes in society's perception and representation of children and/or adults with differences of sex development, which is of great importance.

8 An ethical challenge: moving society's perception towards greater acceptance of differences

This is a fundamental issue when speaking of "different children." For society's perception to change, there is a need **to train, inform, and discuss**. This subject of differences of sex development at birth and their consequences should be included in school curricula (biology classes). More generally, it would be useful to furnish the general public with information on the reality of intersexuality. Society's perceptions of norms and differences are not irreversible, and so the CCNE hopes to see implementation of the measures needed to move such perceptions towards unconditional acceptance of these children in their uniqueness. In this regard, the French national public health agency (Agence Nationale de Santé Publique France) could be commissioned to deliver the appropriate messages.

APPENDICES

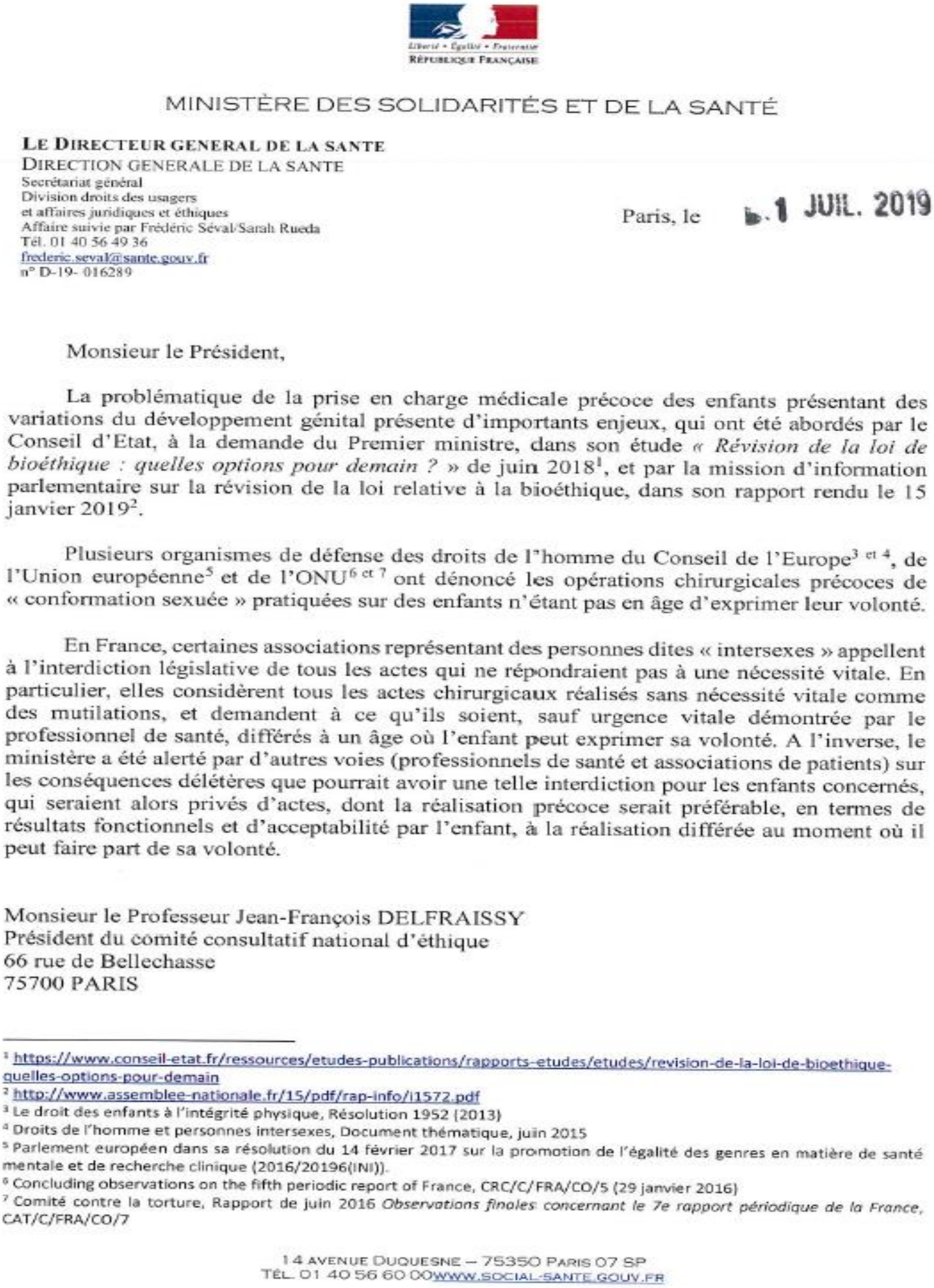
Appendix 1: Members of the working group

Mounira Amor-Guéret
François Ansermet (rapporteur)
Pierre-Henri Duée
Florence Gruat
Karine Lefeuvre
Martine Le Friant
Caroline Martin (rapporteur)
Marion Muller-Colard
Francis Puech (rapporteur)

Appendix 2: Experts consulted

Lise Duranteau and Claire Bouvattier (CHU Kremlin-Bicêtre)
Loé Petit and X (Collectif Intersexes et Allié.e.s)
Martine Cools (Ghent University)
Raphaël Rappaport, Michel Polak, Karinne Gueniche, Yves Aigrain (CHU Necker-Enfants malades)
Pierre Mouriquand (CHU Lyon)
Blaise Meyrat (CHUV Lausanne) and Cynthia Kraus (University of Lausanne)
Vincent Guillot (Organisation Intersex International)
Christine Roujeau, Claudine Colin, Nathalie Colin (Association Surrénales)
Alphératz (linguist)
Anne-Marie Rajon (CHU Toulouse)
Mika Petkova, Benjamin Pitcho, Benjamin Moron-Puech (GISS-Alter Corpus)

Appendix 3: Referral letter



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Paris, le 1 JUIL. 2019

Monsieur le Président,

La problématique de la prise en charge médicale précoce des enfants présentant des variations du développement génital présente d'importants enjeux, qui ont été abordés par le Conseil d'Etat, à la demande du Premier ministre, dans son étude « *Révision de la loi de bioéthique : quelles options pour demain ?* » de juin 2018[1], et par la mission d'information parlementaire sur la révision de la loi relative à la bioéthique, dans son rapport rendu le 15 janvier 2019[2].

Plusieurs organismes de défense des droits de l'homme du Conseil de l'Europe[3 et 4], de l'Union européenne[5] et de l'ONU[6 et 7] ont dénoncé les opérations chirurgicales précoces de « conformation sexuée » pratiquées sur des enfants n'étant pas en âge d'exprimer leur volonté.

En France, certaines associations représentant des personnes dites « intersexes » appellent à l'interdiction législative de tous les actes qui ne répondraient pas à une nécessité vitale. En particulier, elles considèrent tous les actes chirurgicaux réalisés sans nécessité vitale comme des mutilations, et demandent à ce qu'ils soient, sauf urgence vitale démontrée par le professionnel de santé, différés à un âge où l'enfant peut exprimer sa volonté. A l'inverse, le ministère a été alerté par d'autres voies (professionnels de santé et associations de patients) sur les conséquences délétères que pourrait avoir une telle interdiction pour les enfants concernés, qui seraient alors privés d'actes, dont la réalisation précoce serait préférable, en termes de résultats fonctionnels et d'acceptabilité par l'enfant, à la réalisation différée au moment où il peut faire part de sa volonté.

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[1] https://www.conseil-etat.fr/ressources/etudes-publications/rapports-etudes/etudes/revision-de-la-loi-de-bioethique-quelles-options-pour-demain

[2] http://www.assemblee-nationale.fr/15/pdf/rap-info/i1572.pdf

[3] Le droit des enfants à l'intégrité physique, Résolution 1952 (2013)

[4] Droits de l'homme et personnes intersexes, Document thématique, juin 2015

[5] Parlement européen dans sa résolution du 14 février 2017 sur la promotion de l'égalité des genres en matière de santé mentale et de recherche clinique (2016/20196(INI)).

[6] Concluding observations on the fifth periodic report of France, CRC/C/FRA/CO/5 (29 janvier 2016)

[7] Comité contre la torture, Rapport de juin 2016 *Observations finales concernant le 7e rapport périodique de la France*, CAT/C/FRA/CO/7

Afin d'identifier les suites à donner aux rapports et aux sollicitations précités, la direction générale de la santé, avec l'appui de la direction générale de l'offre de soins (DGOS) et de la direction des affaires civiles et du sceau (DACS), a conduit une série d'entretiens avec les parties prenantes, notamment les médecins experts du centre de référence maladies rares relatif au développement génital (CRMR DEV GEN), labellisé par la DGOS en 2017, qui comprend 4 sites principaux et dispose d'un réseau de centres de compétence sur tout le territoire, l'association Amnesty international, l'association Surrénales, le GISS Alter-Corpus et le collectif intersexes et allié.e.s.

A l'issue de ces rencontres avec les principales parties prenantes, l'administration réfléchit à prendre un arrêté sur le fondement de l'article L. 1151-1 du code de la santé publique[8] visant à instaurer un recours systématique de chaque enfant concerné au centre de référence, qui soit assurerait directement sa prise en charge et son suivi, soit en déterminerait les modalités. Cela garantirait que chaque enfant bénéficie d'une démarche diagnostique optimale et que les propositions thérapeutiques émises, y compris d'abstention thérapeutique, reposent sur une concertation d'experts pluridisciplinaires et hautement spécialisés, privilégient, à chaque fois que cela est possible, le report des actes médicaux à un âge où l'enfant peut participer à la décision, et que les enfants et leurs familles bénéficient d'une information complète et d'un accompagnement approprié dès le début de la prise en charge.

Cet arrêté pourrait procéder à une révision du cahier des charges du centre de référence et définir les règles de bonnes pratiques diagnostiques et concernant la réalisation de toute acte altérant les caractéristiques sexuelles des enfants présentant de telles variations du développement génital.

La direction générale de la santé sollicite l'avis du CCNE, qui examine actuellement cette problématique dans le cadre d'une auto-saisine faisant suite à une demande formulée par le GISS, afin de proposer des questions éthiques dont les réponses sont susceptibles d'aider à la rédaction de l'arrêté ou ses documents annexes. Elle s'interroge notamment sur les problématiques suivantes :

- au regard des principes d'intégrité, d'autonomie et d'accès aux soins, et hors urgence vitale, est-il possible de différentier les actes qui peuvent être pratiqués précocement ou être différés, en prenant en compte la diversité des situations, l'objectif de ces actes à court, moyen et long terme ainsi que l'absence de certitude scientifique et de consensus professionnel ? Doit-on considérer que seuls les actes répondant à une nécessité vitale peuvent être pratiqués précocement ou bien que des actes répondant à une nécessité médicale peuvent être justifiés précocement et dans quels buts ?

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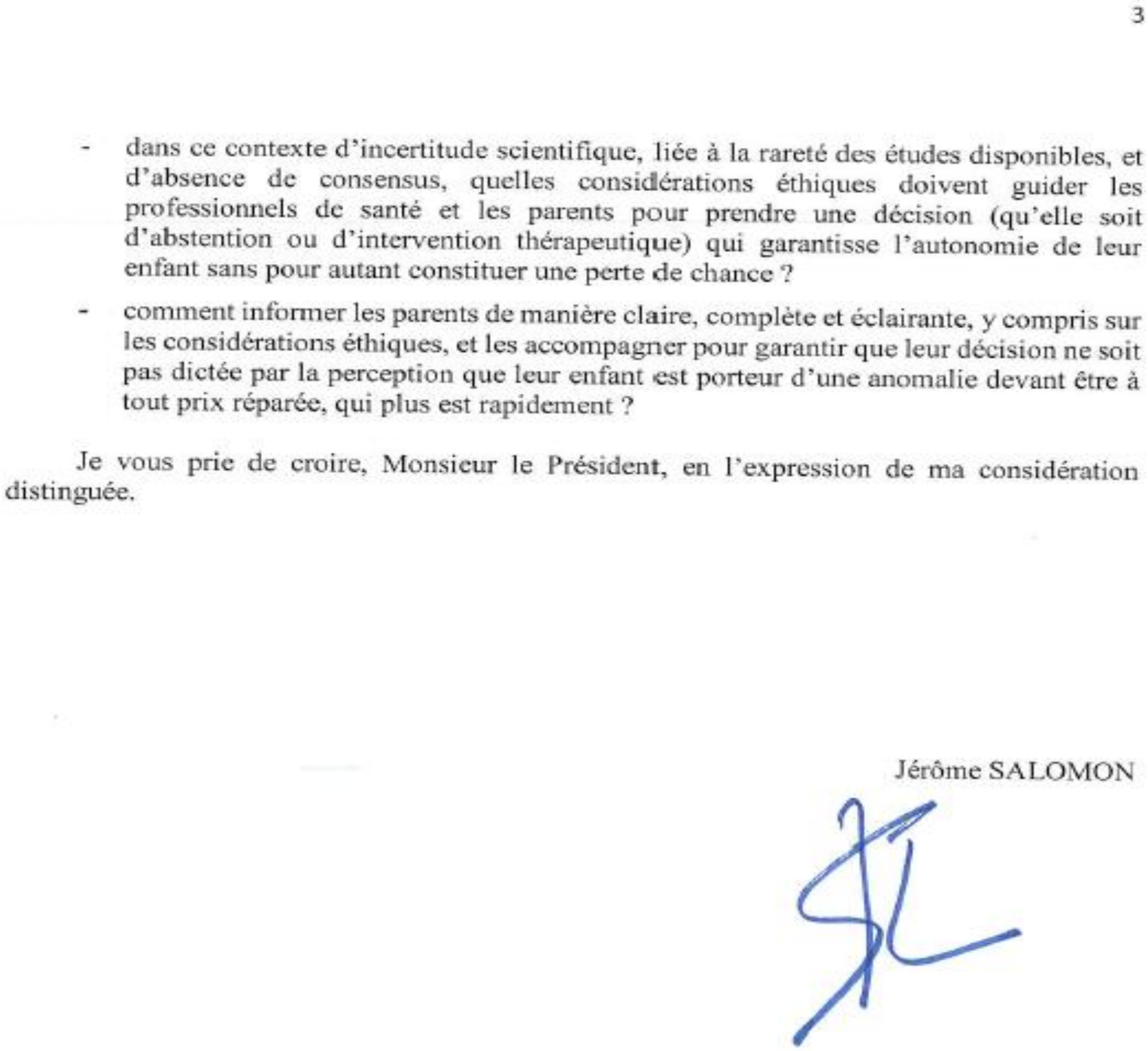
[8] Article L. 1151-1 du code de la santé publique : « *La pratique des actes, procédés, techniques et méthodes à visée diagnostique ou thérapeutique, ainsi que la prescription de certains dispositifs médicaux et l'utilisation de médicaments nécessitant un encadrement spécifique pour des raisons de santé publique ou susceptibles d'entraîner des dépenses injustifiées peuvent être soumises à des règles relatives :*
- à la formation et la qualification des professionnels pouvant les prescrire ou les mettre en œuvre conformément au code de déontologie médicale ;
- aux conditions techniques de leur réalisation.
Elles peuvent également être soumises à des règles de bonne pratique.
Ces règles sont fixées par arrêté conjoint des ministres chargés de la santé et de la sécurité sociale, après avis de la Haute Autorité de santé.
L'utilisation de ces dispositifs médicaux, de ces médicaments et la pratique de ces actes, procédés, techniques et méthodes à visée diagnostique ou thérapeutique peuvent être limitées pendant une période donnée à certains établissements de santé. Les ministres chargés de la santé et de la sécurité sociale arrêtent, après avis de la Haute Autorité de santé, la liste de ces établissements ou précisent les critères au vu desquels les agences régionales de santé fixent cette liste.
Les dispositions du présent article s'entendent sans préjudice des dispositions relatives aux recherches impliquant la personne humaine définies au titre II du présent livre et de celles relatives aux autorisations, aux conditions d'implantation de certaines activités de soins et aux conditions techniques de fonctionnement définies aux chapitres II, III et IV du titre II du livre Ier de la sixième partie. »



- dans ce contexte d'incertitude scientifique, liée à la rareté des études disponibles, et d'absence de consensus, quelles considérations éthiques doivent guider les professionnels de santé et les parents pour prendre une décision (qu'elle soit d'abstention ou d'intervention thérapeutique) qui garantisse l'autonomie de leur enfant sans pour autant constituer une perte de chance ?
- comment informer les parents de manière claire, complète et éclairante, y compris sur les considérations éthiques, et les accompagner pour garantir que leur décision ne soit pas dictée par la perception que leur enfant est porteur d'une anomalie devant être à tout prix réparée, qui plus est rapidement ?

Je vous prie de croire, Monsieur le Président, en l'expression de ma considération distinguée.

Jérôme SALOMON

Dear Mr. President,

The early medical care of children with differences of sex development raises important issues, which were addressed by the Council of State, at the request of the Prime Minister, in its June 2018 study[1] Revision of the Bioethics Law: Which Options for Tomorrow? (in

French) and by the parliamentary information mission on revision of the bioethics law in its report issued on 15 January 2019[2].

Several human rights organizations of the Council of Europe[3 and 4], the European Union[5], and the UN[6 and 7] have denounced early surgical operations of "sexual conformity" performed on children not old enough to express their wishes.

In France, certain advocacy groups representing "intersex" people have called for a legal ban on all procedures when the situation is not life-threatening. In particular, these groups consider all surgical procedures performed when there is not a threat to life as mutilations and demand that these procedures, except when health professionals show there is a medical necessity, be postponed to an age when the children can express their wishes. Conversely, the Minister has been alerted by other channels (health professionals and patient groups) to the deleterious consequences that could arise from such a ban for the children concerned, who would be deprived of procedures early implementation of which would be preferable, in terms of functional results and outcome for the child, to postponement to a time when they can express their wishes.

Professor Jean-François Delfraissy
President of the French National Consultative Ethics Committee
66 rue de Bellechasse
75700 Paris

[1] https://www.conseil-etat.fr/ressources/etudes-publications/rapport-etudes/etudes/revision-de-la-loi-de-bioethique-quelles-options-pour-demain

[2] http://www.assemblee-nationale.fr/15/pdf/rap-info/i1572.pdf

[3] Le droit des enfants à l'intégrité physique, Résolution 1952 (2013)

[4] Droits de l'homme et personnes intersexes, Thematic document, June 2015

[5] European Parliament in its resolution of 14 February 2017 on promoting gender equality in mental health and clinical research (2016/20196(INI))

[6] Concluding observations on the fifth periodic report of France, CRC/C/FRA/CO/5 (29 January 2016)

[7] Comité Contre la Torture, Report of June 2016: Observations finales concernant le 7e rapport périodique en France, CAT/C/FRA/CO/7

In order to identify the follow-up to the reports and to the abovementioned requests, the Direction Générale de la Santé, with the support of the Direction Générale de l'Offre de Soins (DGOS) and of the Direction des Affaires Civiles et du Sceau (DACS), has undertaken a series of interviews with the interested parties, notably the expert physicians of the National Reference Center for Rare Diseases-Genital Development (CRMR DEV GEN), certified by the DGOS in 2017, which comprises 4 main sites and has a network of centers of competence throughout France, Amnesty International, Association Surrénales, GISS Alter-Corpus, and Collectif Intersexes et Allié.e.s.

After these meetings with the main interested parties, the administration considered issuing an order on the basis of article L. 1151-1 of the Public Health Code[8] with a view to implementing systematic referral of each child concerned to the reference center, which will directly ensure the child's care and follow-up, or will determine their modalities. This would guarantee that each child benefits from an optimal diagnostic approach, that the therapeutic proposals made, including non-treatment, based on dialogue between multidisciplinary and highly specialized experts, favor whenever possible the postponement of medical procedures to an age when the child can take part in decision making, and that the family and child receive full information and appropriate support from the outset of care.

This order could lead to a revision of the reference center's specifications and define the rules concerning good diagnostic practice and procedures that alter the sexual characteristics of children with differences of sex development.

The Direction Générale de la Santé seeks the opinion of the CCNE, which is currently examining this problem in the framework of a self-initiated investigation following a request from the GISS, the aim being to raise ethical questions the answers to which are likely to help in the drawing up of the order or its appended documents. The Direction Générale de la Santé is considering in particular the following issues:

- in terms of the principles of integrity, autonomy, and access to care, if the situation is not life-threatening, is it possible to differentiate procedures that can be implemented early from those that can be postponed, taking into account the diversity of situations, the short-, medium-, and long-term aims, and the absence of scientific certainty and professional consensus? Should only those procedures needed to avoid a threat to life be practiced early or should early implementation of such procedures be justified and, if so, for what purposes?

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[8] *Article L. 1151-1 (in French) of the Public Health Code: "The practice of procedures, processes, techniques, and methods with a diagnostic or therapeutic aim, and the prescription of certain medical devices and the use of medications requiring specific management for reasons of public health or likely to lead to unjustified expenses may be subject to rules concerning:*
- the training and qualification of professionals who prescribe or implement them in accordance with the Code of Medical Ethics;
- the technical conditions of their implementation.
They may also be subject to the rules of good practice. These rules are fixed by a joint order from the ministers of health and social security, after consulting the Haute Autorité de Santé.

The use of these medical devices and medications, and the implementation of these procedures, processes, techniques, and methods with a diagnostic or therapeutic aim may be restricted during a given period to certain healthcare establishments. The ministers of health and social security, after consulting the Haute Autorité de Santé, will draw up the list of these establishments or specify the criteria according to which regional health agencies draw up this list.

The provisions of the present article shall be without prejudice to the provisions concerning research involving human subjects defined under heading II of the present book and of those relating to authorizations, to the conditions for the setting up of certain healthcare activities, and to the technical conditions of functioning defined in chapters II, III, and IV under heading II of the 1st book of the sixth part."

- in this context of scientific uncertainty, related to the scarcity of available studies and the lack of consensus, which ethical conditions should guide health professionals and parents in taking a decision (to treat or not to treat) that guarantees the autonomy of the child without constituting a lost opportunity?
- how to provide the parents with clear, full, and enlightening information, including on ethical considerations, and to support them, so as to guarantee that their decision is not dictated by the perception that their child has an anomaly which should, at all costs, be corrected, and quickly?

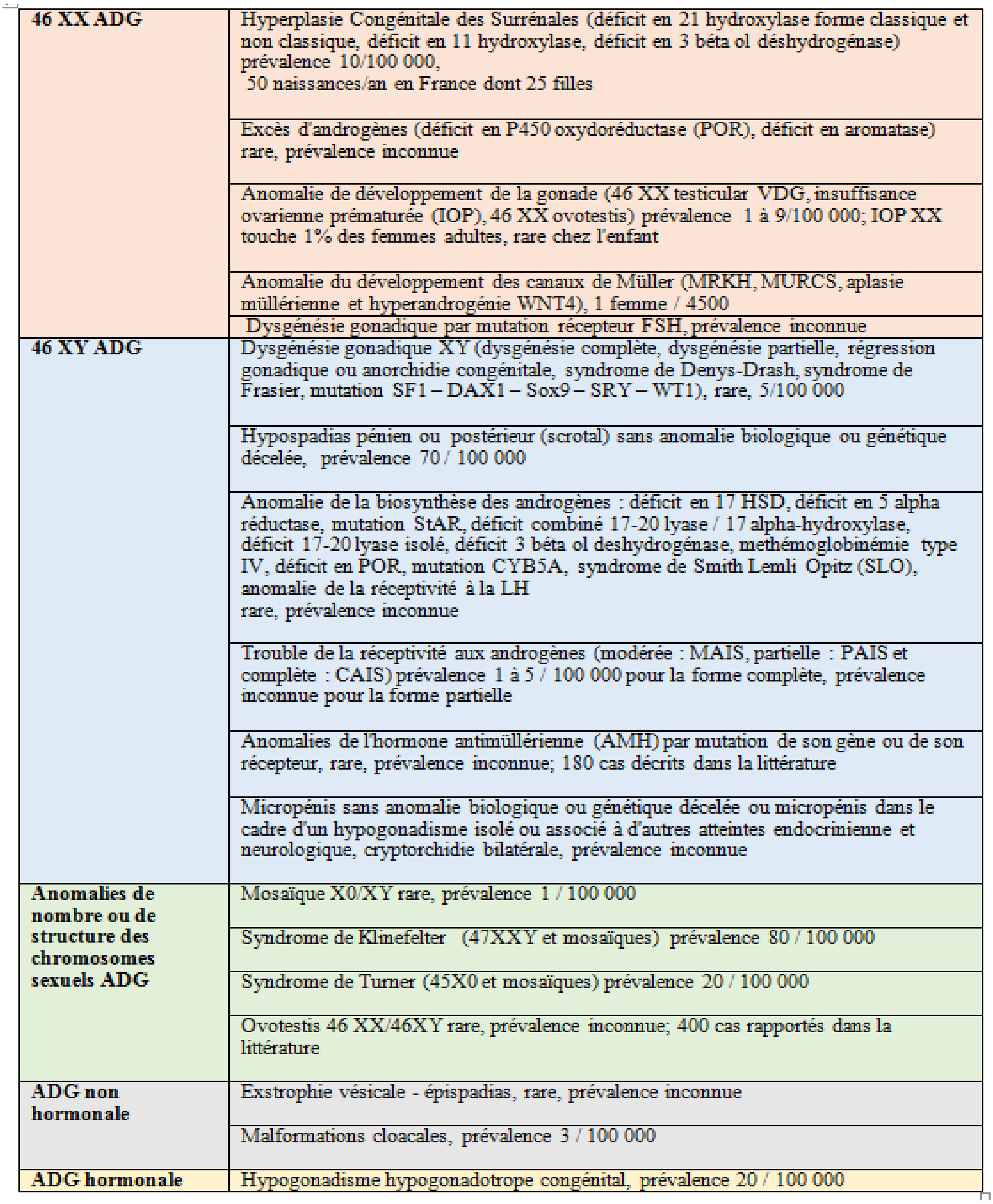
I ask you to accept, Mr. President, the expression of my highest consideration.

Jérôme SALOMON
(signed)

Appendix 4: Detailed list of disorders of sex development (DSD) with, where known, their prevalence per 100 000 population (reference center DEVGen Lille 2017)

46 XX DSD	Congenital adrenal hyperplasia (classic and non-classic 21-hydroxylase deficiency, 11-hydroxylase deficiency, 3-beta-hydroxysteroid dehydrogenase deficiency). Prevalence 10/100 000. 50 births/year in France, including 25 girls
	Androgen excess (P450 oxidoreductase (POR) deficiency, aromatase deficiency). Rare. Prevalence unknown.
	Disorder of gonadal developmental (46 XX testicular DSD, premature ovarian insufficiency (POI), 46 XX ovotestis). Prevalence 1 to 9/100 00; IOP XX affects 1% of adult women; rare in children
	Anomaly in the development of the Müllerian ducts (MRKH syndrome, MURCS association, WNT4 Müllerian aplasia, hyperandrogenism). 1 woman/4500
	Gonadal dysgenesis due to an FSH receptor mutation. Prevalence unknown
46 XY DSD	Gonadal dysgenesis XY (complete dysgenesis, partial dysgenesis, gonadal regression or congenital anorchia, Denys-Drash syndrome, Frasier syndrome, SF1, DAX1, Sox9, SRY, WT1 mutations). Rare. Prevalence 5/100 000
	Penile or posterior (scrotal) hypospadias with no detected genetic anomaly or abnormal laboratory value. Prevalence 70/100 000
	Anomaly in androgen biosynthesis: 17-HSD deficiency, 5-alpha reductase deficiency, StAR mutation, combined 17 alpha-hydroxylase/17,20-lyase deficiency, isolated 17,20-lyase deficiency, 3-beta-hydroxysteroid dehydrogenase deficiency, methemoglobinemia type IV, POR deficiency, CYB5A mutation, Smith-Lemli-Opitz (SLO) syndrome, abnormal LH receptivity. Rare. Prevalence unknown
	Androgen insensitivity syndrome (mild: MAIS, partial: PAIS, and complete: CAIS). Prevalence 1 to 5/100 000 for CAIS; unknown for PAIS
	Anti-Müllerian hormone (AMH) anomalies due to mutation of its gene or of its receptor. Rare. Prevalence unknown. 180 cases described in the literature

	Micropenis without detected genetic anomaly or abnormal laboratory value or micropenis in a setting of isolated hypogonadism or hypogonadism associated with endocrine and neurological involvement, bilateral cryptorchidism. Prevalence unknown
Anomalies in the number or structure of sex chromosomes DSD	X0/XY mosaicism. Rare. Prevalence 1/100 00
	Klinefelter syndrome (47,XXY and mosaicisms). Prevalence 80/100 00
	Turner syndrome (45X0 and mosaicisms). Prevalence 20/100 000
	Ovotestis 46XX/46XY. Rare. Prevalence unknown. 400 cases reported in the literature
Nonhormonal DSD	Bladder exstrophy – epispadias. Rare. Prevalence unknown
	Cloacal malformations. Prevalence 3/100 000
Hormonal DSD	Congenital hypogonadotropic hypogonadism. Prevalence 20/100 000



46 XX ADG	Hyperplasie Congénitale des Surrénales (déficit en 21 hydroxylase forme classique et non classique, déficit en 11 hydroxylase, déficit en 3 béta ol déshydrogénase) prévalence 10/100 000, 50 naissances/an en France dont 25 filles
	Excès d'androgènes (déficit en P450 oxydoréductase (POR), déficit en aromatase) rare, prévalence inconnue
	Anomalie de développement de la gonade (46 XX testicular VDG, insuffisance ovarienne prématurée (IOP), 46 XX ovotestis) prévalence 1 à 9/100 000; IOP XX touche 1% des femmes adultes, rare chez l'enfant
	Anomalie du développement des canaux de Müller (MRKH, MURCS, aplasie müllérienne et hyperandrogénie WNT4), 1 femme / 4500
	Dysgénésie gonadique par mutation récepteur FSH, prévalence inconnue
46 XY ADG	Dysgénésie gonadique XY (dysgénésie complète, dysgénésie partielle, régression gonadique ou anorchidie congénitale, syndrome de Denys-Drash, syndrome de Frasier, mutation SF1 – DAX1 – Sox9 – SRY – WT1), rare, 5/100 000
	Hypospadias pénien ou postérieur (scrotal) sans anomalie biologique ou génétique décelée, prévalence 70 / 100 000
	Anomalie de la biosynthèse des androgènes : déficit en 17 HSD, déficit en 5 alpha réductase, mutation StAR, déficit combiné 17-20 lyase / 17 alpha-hydroxylase, déficit 17-20 lyase isolé, déficit 3 béta ol deshydrogénase, methémoglobinémie type IV, déficit en POR, mutation CYB5A, syndrome de Smith Lemli Opitz (SLO), anomalie de la réceptivité à la LH rare, prévalence inconnue
	Trouble de la réceptivité aux androgènes (modérée : MAIS, partielle : PAIS et complète : CAIS) prévalence 1 à 5 / 100 000 pour la forme complète, prévalence inconnue pour la forme partielle
	Anomalies de l'hormone antimüllérienne (AMH) par mutation de son gène ou de son récepteur, rare, prévalence inconnue; 180 cas décrits dans la littérature
	Micropénis sans anomalie biologique ou génétique décelée ou micropénis dans le cadre d'un hypogonadisme isolé ou associé à d'autres atteintes endocrinienne et neurologique, cryptorchidie bilatérale, prévalence inconnue
Anomalies de nombre ou de structure des chromosomes sexuels ADG	Mosaïque X0/XY rare, prévalence 1 / 100 000
	Syndrome de Klinefelter (47XXY et mosaïques) prévalence 80 / 100 000
	Syndrome de Turner (45X0 et mosaïques) prévalence 20 / 100 000
	Ovotestis 46 XX/46XY rare, prévalence inconnue; 400 cas rapportés dans la littérature
ADG non hormonale	Exstrophie vésicale - épispadias, rare, prévalence inconnue
	Malformations cloacales, prévalence 3 / 100 000
ADG hormonale	Hypogonadisme hypogonadotrope congénital, prévalence 20 / 100 000

AVIS132

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