

useful in the management of individual cases and with the difficult task of maintaining compliance. It also made it possible to identify a small group of children at a very high risk of relapse, essentially young (<5 years of age) non-Caucasian children with severe initial hyperthyroidism. In conclusion, a longer initial duration of a euthyroid state with ATD treatment is the most significant prognostic variable. However, the optimal duration remains to be evaluated in further studies.

Raphaël Rappaport, MD

Second Editor's Comment: Hyperthyroidism is believed to result from a complex interaction between the autoimmune system, environmental factors, and genetic background; it is mainly due to Graves' disease and is less frequently seen in children than in adults. ATD treatment is the initial form of therapy for all hyperthyroid children in an attempt to normalize thyroid function tests. Whether this form of treatment is continued long-term or whether other therapeutic options, such as surgery or radioactive iodine treatment, are considered is often dependant on the rate of relapse after ATD treatment. Reliable predictors of relapse after ATD treatment would facilitate the management of these children by allowing for the identification of those requiring long-term ATD, or alternatively thyroidectomy or radioiodine therapy.¹ In this study, Kaguelidou et al were able to find the 5 variables most predictive of relapse following ATD. At diagnosis the key factors to consider when evaluating the risk of relapse of a patient are: ethnicity (higher risk for children of Non-Caucasian origin), age (the younger the patients the higher the risk for relapse), severity of the disease as manifested by elevated serum free T_4 and TRAb levels (the higher these concentrations, the higher the risk of relapse) and duration of the disease. It is interesting to note that children receiving longer ATD treatment were less likely to relapse, with a 43% decrease in relapse risk for each additional 12 months of treatment. While it is clear that there is no ideal form of therapy for this disease as the 3 available therapeutical options (ATD, thyroidectomy, and radioactive iodine) are associated with potential complications, drug therapy remains the first line of treatment in many countries. The remission rate after 2 years of ATD treatment (about 30%) observed in this study is in agreement with a 1987 report.² This study also demonstrated that the remission rate increases significantly in children and adolescents with every additional year of

treatment. The need for prescribing longer treatment courses in children than in adults is now widely accepted and the duration of medical treatment seems to be the only variable, independent of ethnicity, age and severity of disease, that can be manipulated.

Roberto Lanes, MD

Third Editor's Comment: The value of predictors to determine the relapse risk of patients with hyperthyroidism following ATD therapy has long been studied in children and adults. The most controversial factor is the serum TRAb level which may not be sufficiently sensitive to predict a relapse after ATD treatment³ even though others have considered them useful in children.⁴ TRAb data are often lacking at diagnosis and/or during follow-up of these patients with Graves' disease, as was the case in this study of Kaguelidou et al. However, the long-term results of ATD treatment remain generally unsatisfactory in most studies. Poor compliance with medical therapy is often the most important factor that determines the therapeutic outcome, particularly in adolescents. Yet long-term treatment seems to be the only variable that is at the clinician's control to reduce the risk of relapse of the disease. Thus, important arguments have been put forward for considering ¹³¹Iodine therapy or surgical ablation in the treatment of children with hyperthyroidism.^{5,6}

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Treatment Guidelines for Children with Disorders of Sex Development

Disorders of sex development (DSD) is the umbrella term replacing intersexuality to cover congenital conditions characterized by atypical chromosomal, gonadal, or anatomic sex.¹ This article was published in a special issue of a journal focusing on gender identity disorders (GID). However, Meyer-Bahlburg

sees sufficient differences between gender-variant persons, with and without a DSD, to urge distinct evaluation and treatment approaches.

GID is characterized by discomfort or distress with one's apparent or assigned gender accompanied by a persistent identification with the opposite sex. In

contrast, gender issues may be far less salient for those with DSD. The challenges related to having a chronic (for some, a life-threatening) medical condition, and its associated management, require that the behavioral health professional be competent in the application of psychosocial interventions for problems of medical adherence and in coping with the stigma often associated with congenital or chronic conditions, in general.

Whereas the evaluation and psychosocial treatment of persons with GID can be conducted by a mental health provider alone, the 2005 consensus statement on DSD¹ calls for care to be provided in the context of a multidisciplinary team, including: neonatology, pediatric endocrinology, pediatric urology, gynecology, genetics, genetic counseling, mental health specialists, social work, nursing, and medical ethics. Moreover, communication between the DSD team and the “medical home” (ie, the primary care physician) is strongly recommended. Because of the high stress at the time of ascertainment of the DSD, there is a critical need for good communication among team members and the family to facilitate shared decision making regarding gender assignment and surgical options. Information shared with the family must include the most up-to-date information regarding the patient’s specific syndrome, including the range of prognostic outcomes for both physical health and psychological health across the lifespan.

Meyer-Bahlburg describes in considerable detail the complexities of the process of gender assignment, and potential reassignment, for those diagnosed at birth and the long-term risk for the child and family associated with missteps in clinical management. Providing integrated interdisciplinary team care for patients is complex and time-consuming for patients, in general. In the case of DSD, Meyer-Bahlburg notes the importance of sustaining the team approach beyond the period of initial diagnosis and early interventions. Optimal care also requires active outreach by the behavioral health provider of the team to adopt a preventive approach regarding problems with psychosocial adaptation and medical adherence. Children and their families may require assistance in interpreting gender-atypical behavior – a not infrequent occurrence in DSD – as understandable based on what is known of the biology of DSD, rather than as a sign of incorrect gender assignment.

Young children with a DSD, who were misdiagnosed or late diagnosed, create special challenges. Parents may require reassurance and counseling if gender atypical behavior is part of the presentation. Gender reassignment after infancy requires careful psychological evaluation over a prolonged period with particular attention to the child’s gender-role behavior and to any symptoms of gender dysphoria.

There is no controversy over performing genital surgery for acute medical reasons. However, medical urgency is the exception rather than the rule. Instead, genital surgery has been performed, typically early in life, to “confirm

the assigned gender by genital appearance.” At present, there is no broad consensus regarding the issue of early genital surgery, with the exception of the milder cases of atypical genital development for which deferring surgery is now recommended.¹ Meyer-Bahlburg describes the preparation required for parents and, later, patients regarding genital surgery decisions. The psychological risks for the patient associated with repeated genital examinations, in part to support the training of medical students and residents, requires a rethinking of medical educational models and practices.

This article provides additional guidance regarding androgen treatment in 46,XY children with underdeveloped genitalia and the timing of sex hormone treatment in persons without gonads or with under-functioning gonads. In general, the timing of hormone replacement is best initiated during the period when peers are experiencing endogenous puberty.

The topic of disclosure of medical information to the patient is a crucial component of psychosocial management. Although legal standards generally support the rights of parents to determine what and when details of their child’s medical condition is disclosed to them, the majority of clinicians agree that a patient with a DSD be fully informed of all details. Some parents will resist disclosing information to their child or adolescent, in particular in cases in which the assigned gender is at odds with sex chromosomes or gonadal structure. Parents can be reassured by the experience of patients with other medical conditions (eg, pediatric cancer or HIV) that disclosure is associated with enhanced psychosocial adaptation. Meyer-Bahlburg reviews strategies for the disclosure process, including providing a web link to a source for animated visual aids.

Finally, the beneficial role of support groups for persons with medical conditions, in particular for those with rare conditions, is emphasized. Health care providers are encouraged to seek opportunities to dialogue with such groups to ensure that the information disseminated is accurate.

Meyer-Bahlburg HFL. Treatment guidelines for children with disorders of sex development. *Neuropsychiatrie de l’Enfance et de l’Adolescence* 2008; 56:345-349.

Editor’s Comment: *Important distinctions between persons with GID and DSD are often blurred in both the popular and scientific literature. While the entities may share some features (eg, gender concerns), DSD diverge from GID in terms of associated features including prevalence, age of onset, and sex ratio.² Failure to differentiate between persons with and without a clearly identifiable DSD may hamper studies of etiology and optimal clinical management.*

The recently published consensus on the management of DSD¹ is very clear on the necessity of applying an integrated interdisciplinary team approach to the care of those affected and their families. What

is not discussed are the barriers that exist to forming and sustaining such teams. The non-reimbursable time required to organize a team is substantial and the essential behavioral health component of service is frequently excluded because clinical services provided by mental health providers are often carved-out by many health insurance plans and require the patient to be seen by an approved insurance panel member who would not be a member of the DSD team. Co-pays for mental health services are characteristically substantially higher than for medical services, forcing families to reject recommended services delivered by the behavioral health member of the team. At present, there is a scarcity of behavioral health experts qualified to immediately join emerging DSD teams. However, the skills set of pediatric psychology makes this subspecialty of child clinical psychology ideally suited to serve as team members at centers of excellence called for in the DSD consensus statement.¹

An evidence-based consensus on best clinical practices regarding gender assignment and genital surgery is only beginning to emerge; in the meantime

there is critical need for systematic investigation to understand how parents are counseled and select among treatment options. Clinicians and representatives of patient advocacy organizations voice concerns about the extent and quality of information disclosed to parents during the decision-making process and, importantly, the subsequent validity of parental consent. These factors make this an excellent clinical context in which to study parental medical decision-making.

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