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Clinical Management of Menstruation in Adolescent Females With Developmental Delay

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Abstract: We review the main concerns related to the management of menstruation in adolescent females with developmental delay. Results from a questionnaire evaluating the current practice of menstrual control in developmentally delayed females by pediatric endocrinologists are also summarized. Guidelines for clinical care are provided, taking into account the specific health issues of this particular patient population.

Key Words: menstruation control, developmental delay, contraception

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Learning Objectives:

- Identify the problems that may confront developmentally delayed adolescent girls, focusing on sexuality, adverse emotional and behavioral changes, and the timing of puberty.
- Compare methods of menstrual control and contraception that are currently considered for developmentally impaired adolescents.
- Appraise guidelines for specific and practical measures that may be taken to optimize menstrual function and contraception in postpubertal girls who are developmentally delayed.

An adolescent girl with developmental delay presents a unique set of reproductive health concerns to both the physician and caregiver.¹ Advancements in chronic health

care for the developmentally disabled population have led to an increased emphasis on improved socialization and less institutionalization for these patients. Physicians are faced more often with issues related to menstrual control in girls with developmental delay.^{2–4} Although several publications have addressed contraceptive management in women with intellectual disability, review of the available literature revealed a lack of specific guidelines addressing the unique problems related to menstruation in this particular group of adolescents. Moreover, responses to a questionnaire sent to pediatric endocrinologists, to assess the current practice of menstrual control in these girls, indicated great variability in management approach. This article gives an overview of the major clinical and reproductive concerns affecting adolescent females with developmental delay. Special attention is given to contraceptive management of menstruation, and practical guidelines for clinical care are provided.

CLINICAL AND REPRODUCTIVE CONCERNS

Personal Hygiene

Developmentally delayed adolescents could be unable to cooperate with daily standard hygiene routines. Associated problems such as relative immobility, coexisting contractures, or behavioral difficulties could prevent normal participation in personal hygiene. Bladder or bowel incontinence further complicates the self-care required during menses, which could be particularly problematic in girls with severe cognitive impairment.⁵

Sexuality

The timing of puberty could be normal, early, or delayed in adolescents with developmental delay. Sex steroid production is accompanied by sexual feelings similar to those experienced by normal adolescents.⁶ For those girls able to willfully express their sexuality, appropriate socialization and sexual expression must be supported.⁷ For those without decisional capacity, the approach to care must focus on protection from sexual abuse. Sexuality education should

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begin late in childhood/early in adolescence to be effective. In patients who lack the ability to understand their sexuality, different modes of intervention are indicated to avoid future inappropriate behaviors.⁸

Susceptibility to Sexual Abuse

Of particular concern for parents of girls with developmental delay is the potential for sexual abuse.¹ A young woman with apparently mild cognitive impairment could lack the capacity to give informed consent for sexual interaction. However, she could frequently be in social situations where unwanted sexual contact could occur. Girls with more severe cognitive disabilities might not be able to voice concern at all and might not protest or report that inappropriate sexual activity is occurring. Openness to the discussion of these issues by all healthcare providers is critical to ensure prevention of abuse and to provide timely contraception.

Effects of Cyclical Hormonal Changes

Several studies have shown an increase in seizure frequency at certain stages of the menstrual cycle.⁹ Catamenial seizure exacerbation has been related to changes in progesterone concentration.¹⁰ Cyclical hormonal changes have also been shown to lead to adverse emotional and physical sequelae as part of the premenstrual syndrome. A retrospective chart analysis documented that 18% of adult females with mental disability had cyclical behavioral changes that could be attributed to the hormonal changes associated with the premenstrual syndrome.¹¹ This compares with only 5% in the general female population.¹² Behavioral changes can be related to menstrual cramping and pain, and these changes are different from those found during the premenstrual syndrome in women without developmental disability.¹¹ Other behavioral symptoms seen in women with developmental disability include aggression, restlessness, hyperactivity, increased agitation, and self-mutilation. Favorable treatment responses are observed in women treated with nonsteroidal antiinflammatory drugs or hormonal manipulation of the menstrual cycle.¹³

Abnormal Timing of Puberty

Precocious puberty is 20 times more common among individuals with neurodevelopmental disabilities than in normal children, which is not surprising given their high rate of central nervous system abnormalities.¹⁴ Central precocious puberty can be treated with gonadotropin-releasing hormone-analog therapy. In the child with severe cognitive impairment, it is important to determine whether final adult height is a priority when considering therapy to suppress early pubertal changes. Permitting or fostering relative short stature could be actually desirable in severely delayed patients.

Delayed onset of puberty could also occur in girls with central nervous system anomalies. Although the caregivers could encounter relief from this delayed pubertal develop-

ment, it can further aggravate preexisting impairment in bone mineralization.¹⁵ In addition, it is well known that anticonvulsant therapy can have a negative effect on bone health, putting those girls with associated seizure disorder at an even higher risk for early-onset osteoporosis, with concurrent increased risk for fractures.¹⁶

CURRENT PRACTICE OF MENSTRUAL CONTROL AMONG PEDIATRIC ENDOCRINOLOGISTS

In 2003, the Drug and Therapeutics Committee of the Lawson Wilkins Pediatric Endocrine Society used a questionnaire to assess the current practice of menstrual control of adolescent females with developmental delay with practicing pediatric endocrinologists in the United States and Canada. Eighty-four percent of (a total of 100) respondents indicated that they regularly receive consults for menstrual control in this patient population. Of all pediatric endocrinologists dealing with this issue, 95% receive less than 10 consults annually for this indication. Specific reasons for obtaining a consultation were: hygiene issues (26%), fear of pregnancy (19%), fear of sexual abuse (16%), dysfunctional uterine bleeding (10%), precocious puberty (22%), specific sterilization request (5%), psychosocial concerns (1%), and other (1%; "inconvenience" and osteoporosis risk). On the other hand, 25% of respondents felt uncomfortable treating adolescent females with developmental delay for menstrual cycle control. Nearly one third of pediatric endocrinologists will not manage adolescent females with developmental delay long-term, preferring to refer to a gynecologist. The different methods used by pediatric endocrinologists to address the problems with menstrual control in developmentally delayed girls are summarized in Figure 1.

CONTRACEPTIVE MANAGEMENT

Developmentally delayed girls have specific needs for their contraceptive management, depending on the mental (and sometimes physical) capabilities of the individual patient.¹⁷ Contraception is considered to stop menses—to facilitate overall hygiene and personal care, prevent unwanted pregnancy, and suppress hormonal changes that could exacerbate certain behavioral difficulties or interfere with seizure control. Finally, adolescents with developmental delay could become sexually active and appropriate reproductive health care must be part of their treatment. Physicians could be uncomfortable with issues related to sexuality and contraception, because they might not have been trained to address these, because reproductive healthcare needs of the mentally disabled have received less attention in the past. Training in the contraceptive management of these developmentally delayed females should be provided as part of residency training in pediatrics, family practice, internal medicine, and gynecology.

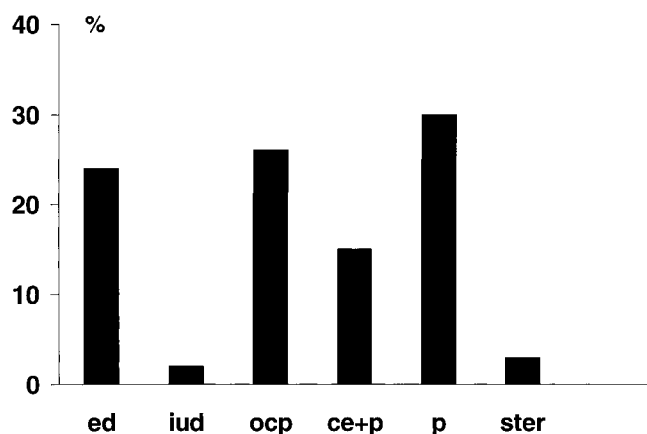


FIGURE 1. Methods used by pediatric endocrinologists to treat developmentally delayed females for concerns regarding menstruation control. ed, education and counseling; IUD, barrier methods and IUDs; ocp, combination estrogen and progestin with monthly cycling; ce+p, continuous estrogen with progestin withdrawal every 3, 4, or 6 months; p, progestin-only therapy; ster, endometrial ablation or hysterectomy.

Barrier methods might not be very practical for individuals with significant cognitive (and/or physical) disabilities.^{3,17,18} Barrier methods require a high degree of personal initiative, intellectual understanding, and physical dexterity. Patients with cognitive impairment are rarely able to use these devices reliably.

Intrauterine devices (IUDs) are usually not recommended for patients with cognitive impairment. Patients are often unable to report pain or discomfort that could accompany a medical complication resulting from the insertion of an IUD. IUDs could lead to menorrhagia and dysmenorrhea, increasing menstrual hygiene problems.

Oral contraception is commonly used in patients with cognitive impairment. The associated decrease in menstrual flow with oral contraceptive use is considered of great benefit. Although oral contraceptives are very effective and of relatively low risk to most patients, they might not always be the best choice if compliance is a concern. The risk of thromboembolism must also be kept in mind, especially for immobile patients. The dose of oral contraceptives may need to be adjusted if the patient is also on anticonvulsants.

Long-acting progestins are another reliable method of contraception,¹⁹ associated with easier hygienic care because of the concomitant amenorrhea. However, potential adverse effects from medications such as depo-medroxyprogesterone acetate (DMPA) or levonorgestrel implants include mood and behavioral changes, weight gain, pain from injection, fatigue, menstrual pattern irregularity, and estrogen deficiency resulting in impaired bone mineral accretion.

LEGAL AND ETHICAL ISSUES

Discussions of legal and ethical issues surrounding menstrual cycling and reproduction in developmentally delayed females have focused primarily on sterilization (endometrial ablation, hysterectomy). In the early 1900s, forced sterilization of girls and women with developmental disabilities was encouraged and even required by certain state laws.²⁰ In 1942, the U.S. Supreme Court declared human procreation a fundamental right, leading to changes in the legality of sterilization of those deemed mentally incompetent.²¹ Sterilization became increasingly difficult or prohibited in some jurisdictions. In the late 1970s, U.S. regulations prevented the use of federal funding to sterilize individuals with developmental disability. During the last 2 decades, additional ethical guidelines have placed further limits on sterilization of individuals with developmental disabilities.^{21–23}

In 1999, the American Academy of Pediatrics (AAP) Committee on Bioethics published recommendations for use when considering sterilization of minors with developmental disabilities.²¹ The recommendations state that consideration of sterilization should focus on the goals of those requesting it, to differentiate between those who want permanent prevention of pregnancy from those who want to avoid other consequences of sexual maturation (for which there could be other options). Frequently requests for sterilization of persons with developmental disability are made to avoid these latter problems, which can be dealt with using different, less intrusive interventions such as developmentally appropriate education with or without medical treatment. Sterilization of a minor must now be approved by the court after a thorough legal and ethical review of each case. It is now difficult to obtain approval for sterilization for a young woman if she demonstrates enough comprehension and competence to raise the possibility of maintaining a long-term relationship in the future or of becoming part of a family. In Canada, sterilization of a mentally disabled woman is illegal.

Although somewhat outside the scope of this article, attention needs to be drawn toward the relative cost and insurance coverage (or lack thereof) of birth control. Many health insurance plans in the United States do not provide coverage for birth control pills and other contraceptives. Out-of-pocket expenses for these agents easily add up to \$300 to \$500 per year. Because federal legislation on this matter has been stalled in Congress for several years, some states have passed laws requiring health plans to pay for contraceptives, and several other states are considering such legislation. Nevertheless, a physician prescribing contraceptives for menstrual control in a mentally disabled female might need to provide documentation of “medical necessity” for the patient to obtain payment from her health insurance company.

GUIDELINES FOR CLINICAL CARE

Manipulating the menstrual cycle in adolescent females with developmental delay is relatively easy. Nevertheless, certain factors in the patient's medical history and physical examination should be considered when opting for contraceptive medication in these girls. Both absolute and relative contraindications to the use of contraceptives will need to be reviewed with the caregivers. The requirement of informed consent can be difficult for those patients whose mental capacity does not allow them to provide assent. Parents and caregivers can obtain recommendations from certain advocacy groups for the mentally disabled (eg, *The Arc of the United States*). Treatment should be considered if it benefits the patient, not if done solely for the benefit of the caregiver.

Estrogen/progestin combination contraceptives (see Table 1) control the menstrual cycle through a variety of mechanisms, but mainly by suppressing ovulation. Continuous oral monophasic low-dose estrogen/progestin is an appropriate method of contraceptive treatment of adolescent females with developmental delay. If low enough doses of estrogen are being used, there is essentially no need for

monthly withdrawal bleeding.²⁴ Patients take the "active" tablets but skip the placebo pills. A progestin withdrawal can be provided every 4 to 6 months if necessary to prevent spotting. Often, patients will need to transition first through a pattern of withdrawal bleedings every 7 to 10 weeks to achieve amenorrhea. The use of weekly patches containing ethinyl estradiol combined with a progestin (Table 1) can also be used continuously, for example, 9 patches applied serially followed by a 7-day patch-free period to achieve withdrawal bleeding. When contraception is the sole indication for medication use, some of the newer estrogen/progestin combinations administered by intramuscular injection (Table 1) provide more convenience and better compliance.²⁵

If there is development of or worsening of headaches and/or behavioral changes associated with menses, oral contraceptive agents should not necessarily be discontinued. Continuous transdermal estrogen/progestin could help reduce these symptoms. Alternatively, a formulation containing the traditional 21 days of active pills, followed by only 2 days of placebo pills and then 5 days of pills containing only 10 μ g ethinyl estradiol, could be of benefit in reducing symptoms

TABLE 1. More Commonly Used Agents for Contraceptive Management*

Preparation	Remarks
Combined (estrogen and progestin) oral contraceptives	
Alesse, Levlite	20 mcg EE/0.1 mg levonorgestrel
Loestrin Fe 1/20	20 mcg EE/1 mg norethindrone acetate 75 mg ferrous fumarate (inactive tablets)
Mircette	20 μ g EE/0.15 desogestrel 10 μ g EE (last 5 out of 7 "placebo" pills)
Lo/Ovral	30 μ g EE/0.3 mg norgestrel
Desogen	30 μ g EE/0.15 mg desogestrel
Yasmin	30 μ g EE/3.0 mg drospirenone (antimineralocorticoid property)
Ortho-Cyclen	35 μ g EE/0.25 norgestimate
Demulen 1/35	35 μ g EE/1 mg ethynodiol diacetate
Ortho-Novum 1/35	35 μ g EE/1 mg norethindrone
Cyclessa	25 mcg EE/0.1, 0.125, and 0.15 mg desogestrel (triphasic regimen)
Seasonale	30 μ g EE/0.15 levonorgestrel (84 days extended regimen + 7 placebo days)
Progestin-only oral contraceptives	
Micronor, Nor-QD	0.35 mg norethindrone
Ovrette	0.075 mg norgestrel
Patches—weekly	
Ortho-Evra	Delivers 20 μ g EE + 0.15 mg norelgestromin daily (worn for 1 week for 3 consecutive weeks, followed by a fourth week which is patch-free)
Injections	
Lunelle/Cyclo-Provera	25 mg medroxyprogesterone acetate and 5 mg estradiol cypionate intramuscularly every 28 days
Depo-Provera	150 mg depot medroxyprogesterone acetate intramuscularly every 11–13 weeks

*The list is not all-inclusive, but provides a quick reference to some of the contraceptive agents available in the United States. Products containing 50 μ g EE were omitted because they are rarely prescribed. For the oral contraceptives, only 28-day regimen products were included.
EE, ethinyl estradiol.

related to estrogen withdrawal. In case of associated neurovascular headaches, or with continuous headaches not associated with menses, the use of a progestin-only method is recommended.

Depo-medroxyprogesterone acetate (Table 1) could still be the best method for females on anticonvulsant drugs. DMPA could actually decrease the seizure frequency, and the effectiveness of anticonvulsant therapy will not be compromised.²⁶ Otherwise, when patients are on hepatic enzyme-inducing agents such as many anticonvulsants, a 30- to 35- to 50- μ g ethinyl estradiol pill with reduced pill-free interval will have to be used. Adverse effects with the use of DMPA include irregular menstrual bleeding, weight gain, and osteopenia (see subsequently in this article) in the adult female population. The incidence of irregular bleeding is 30% in the first year and 10% thereafter. Irregular bleeding appears to be less frequent in younger teenagers who are often already anovulatory. The injectable estrogen/progestin combinations are an alternative if irregular breakthrough bleeding is a problem. Patients who need to avoid estrogens such as immobile patients and other patients at increased risk for venous thromboembolism are also candidates for DMPA.

When a progestin-only medication is used, the long-term impact on bone mineralization is not well known. Some preliminary data from ongoing studies suggest that there could be mineralization suppression (by DMPA) during adolescence. Supplemental calcium plus vitamin D is therefore advisable. Optimally, prophylaxis with vitamin D (800 IU per day) and supplemental calcium should already be part of the medical management of anticonvulsant-treated patients.

In conclusion, for adolescent females with mild developmental delay, combined estrogen/progestin therapy seems most appropriate. The monophasic pills with low-dose estrogen can be given continuously without need for withdrawal bleeding in most patients. If menstrual control is desirable with monthly menses, newer formulations using estrogen/progestin patches or monthly injections of estrogen/progestin could be the most convenient methods. For girls with more severe mental impairment, the goal for therapy should be to decrease the frequency of or eliminate menstrual cycles while providing adequate contraception. This can be achieved with continuous estrogen/progestin or progestin-only treatment.

With improvement in quality of life for mentally retarded patients, and changes in societal attitude, these individuals are enjoying more normal and satisfying sexual experiences. Associated with these changes are increased demands on physicians to advise these patients and their families about sexual activity as well as contraceptive methods. Physicians should feel comfortable recommending contraception to females with developmental disabilities. The most practical and convenient regimen should be recom-

mended, taking into consideration the individual needs of each patient. Further clinical studies should be helpful in increasing our knowledge regarding the (potential adverse) effects of menstrual cycle manipulation in developmentally delayed females.

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