

regression, approximately a quarter of this group will lose this ability. Overall, approximately 60% remain ambulatory.[25] Toe walking is common, as well as abnormal foot postures in which case ankle/foot orthoses or other orthotic devices should be implemented. Significant effort should be allocated to maintaining ambulation as long as possible. In those girls who do not walk, standing frames should be employed both at home and at school along with parallel bars to promote supported ambulation.

#### ○ Sexual Maturation & Gynecologic Issues

As adolescence approaches, puberty should be anticipated. Ordinarily, girls with RS will enter puberty at similar ages to their peers. As these young women are quite vulnerable, appropriate consideration should be given to protecting them from unwarranted contact.

Menstrual cycles generally occur with predictable regularity after they have become established. Parents or other responsible caregivers may choose to deal with these in the usual fashion or may choose one of a variety of strategies to eliminate or minimize menstrual flow. These include Depo-Provera, birth-control pills, or endometrial ablation. Depo-Provera is not recommended, primarily because it adversely affects bone mineralization, but also because of its long period of action should other side effects arise. The use of birth-control pills is common and completely acceptable. Formulations of levonorgestrel/ethinyl estradiol provide menstrual management for 1 year (marketed as Lybrel®) or for 90 days (marketed as Seasonale® or Seasonique®), such that menstrual flow can be effectively eliminated or minimized.

When unexplained irritability or inconsolable crying occurs, consideration should be given to a possible gynecologic issue. An abdominal ultrasound, as well as consultation with a gynecologist, should be considered strongly.

#### ○ Cardiac & Autonomic Systems

A prolonged QT interval and ST-segment changes may be observed with increasing frequency through the age range.[30,32] As such, at the time of diagnosis, an ECG is recommended to assess QTc or other significant abnormality. If normal, repeating the cardiogram in 2-3 years is appropriate. If abnormal, referral to a cardiologist for consideration of an appropriate intervention strategy is indicated. Prolonged QTc is a quite treatable problem with medication and should not be overlooked. Should a QT-segment abnormality be identified, it would be appropriate to have other members of the patient's family screened in the event this finding is unrelated to the RS.

The hands and feet tend to be cool to cold in RS. This is more typically noted in the lower extremities and may consist not only of cold but discolored feet, ranging from red and mottled to purple, and extending well up on the lower extremity. This finding is believed to be related to excess sympathetic nervous system tone. No specific intervention is available, but it is important to keep the hands and feet appropriately warm.

#### ○ Bruxism

Bruxism (teeth grinding) occurs in virtually all girls at some point in time, varying in frequency and intensity, and exacerbated by anxiety, excitement or stressful situations. The sound created is really quite characteristic, having been described by Bengt Hagberg as that of slowly uncorking a bottle of wine.

Efforts to reduce the teeth grinding are generally unrewarding. Over time, they diminish, in many actually disappearing after school age.

#### ○ Other Motor System Functions

Hypotonia is the rule early in RS. Over time, muscle tone may be increased, representing rigidity rather

# Talking About Psychiatry

**DJ Losen**



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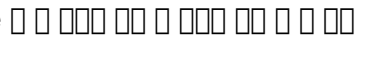
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