Clicker Test

• Which of the following statements best applies to your experience with PAG?
  A. Expert: I could give the lecture
  B. Experienced: I regularly see adolescents in my practice
  C. Eager to learn: I would like to see more adolescents in my practice
  D. Inexperienced: What’s PAG? And isn’t this supposed to be the session on LARC?

GROWTH ATTENUATION THERAPY IN GIRLS WITH DISABILITIES: A DEBATE OF THE ASHLEY TREATMENT

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Disclosures

• Susan Ernst, MD has no disclosures
• Paula Hillard, MD has no disclosures

We will discuss off label use of hormonal medications used for growth attenuation therapy

OBJECTIVES—ASHLEY TREATMENT

At the conclusion of this presentation, the participant will be able to:
1. Outline medical options for growth attenuation for girls with severe developmental delay
2. Discuss the ethical positions argued for and against various components of the Ashley treatment including medical therapy for growth attenuation as well as surgical intervention with mastectomy and hysterectomy.
3. Identify educational interventions as well as social support for families of children with severe disabilities as an alternative to growth attenuation.
Who was Ashley?

- ASHLEY: 6yo Static encephalopathy, G tube dependent multiple impairments unable to sit or stand unassisted, nonverbal
- Evidence of early puberty, parents requested growth attenuation treatment so that Ashley would be easier to care for and remain living with her family
- Ethics committee approved
  - High dose estrogen therapy to close growth plates
  - Removal of breast buds
  - Hysterectomy
  - Appendectomy

Gunther and Diekema: Arch Ped Adoles Med 2006;160: 1013-1017

What May Be Asked of Physicians?

- Growth Attenuation
- Hysterectomy/Sterilization
- Mastectomy
What May Be Asked of Physicians?

- Growth Attenuation
- Hysterectomy/Sterilization
- Mastectomy

Growth Attenuation Therapy (GAT) for Girls with Severe Physical and Cognitive Disabilities (SPCD)

- Before this lecture had you heard of the Ashley case and the debate on GAT?
  - 1. YES
  - 2. NO
Medical Issues of Growth Attenuation

• Historical Perspective:
  • Use of high dose estrogens to limit height in “excessively tall girls”
  • No published series on regimens or efficacy or side effects or risks/benefits in a population with developmental disabilities

• Efficacy and Hormonal Regimens
Medical Issues of Growth Attenuation

• Medical Risks
  • High Dose Estrogen Therapy
    • Risk of VTE
    • Risk of Breast Cancer
  • Unopposed Estrogen and Effect on Endometrium/Bleeding
  • Increased risk of melanoma
    • Cohort study Swedish tall girls, treated vs untreated (Benyi, 2014)
Medical Issues of Growth Attenuation

• Endocrinology /Gynecology (PAG vs Adult)
  • Who sees these girls?
  • Who is best equipped to address needs of girls and parents?

Growth Attenuation Therapy (GAT) for Girls with Severe Physical and Cognitive Disabilities (SPCD)

• Have you been asked to prescribe GAT for girl(s) with SPCD?
  • 1. YES
  • 2. NO
Growth Attenuation Therapy (GAT) for Girls with Severe Physical and Cognitive Disabilities (SPCD)

- Regardless of whether you have previously been asked to provide GAT, do you believe that GAT is a reasonable treatment option for girls with SPCD?
  - 1. YES
  - 2. NO

Which children can be considered for growth attenuation therapy?

- Children with persistent, profound impairment including those with severe developmental and intellectual disability IQ <20 - 25
- Children requiring assistance with all activities of daily living (ADLs) such as feeding, dressing, toileting, mobility

Wilford et al. Hasting Center Report November 2010
Ethical Points of Agreement re Growth Attenuation Therapy

• Shared viewpoints:
  • We are concerned that many people and institutions in society do not positively value people with profound developmental disabilities
  • We agree that investing in improvements in medical and social services is a priority
  • We agree that parents of children with profound developmental disabilities face a complicated set of challenges and should be afforded respect and considerable deference in making the complex and difficult decisions unique to their child’s care


Ethical Arguments in Favor of Growth Attenuation

• Growth attenuation facilitates caregiving
  • Lifting, turning, carrying, transferring
• Growth attenuation can contribute to a child’s quality of life, as opportunities for familial, social, and recreational activity may be limited by size and mobility
  • Travel, beach, park where lifts are not available
Ethical Arguments in Favor of Growth Attenuation

• Parents are usually given discretion in foregoing other treatments that may limit growth:
  • Allowing precocious puberty to progress
  • Refusing g-tube
  • Refusing CPAP
  • Refusing T&A for sleep apnea

Ethical Arguments in Favor of Growth Attenuation

• While GAT may benefit Ashley’s parents, typically the interests of children and parents coincide and are often intertwined

• Goals of parenting include maintaining the child’s current well-being (as opposed to future well-being)
Ethical Arguments Against Growth Attenuation

• GA Devalues people with Disabilities
• A problem with Social Justice
• GA requires changing the body of person with disability instead of changing society to meet the needs of the disabled
• Keeping a child small for ease of care benefits caregiver/society not the child


Ethical Arguments Against Growth Attenuation

• GA Impacts Social Resources
• GA competes with the alternative strategy of improving social services for people with disabilities
• If GA is seen as a cheaper solution, its existence may compound the problem of inadequate resources and services for this population
Ethical Arguments Against Growth Attenuation

- GA may be misused
- GA might be seen as beneficial in other populations of kids with disabilities – those with severe behavioral problems (autism) or those with severe physical disabilities but not cognitive impairment

Ethical Arguments Against Growth Attenuation

- GA may not be in the child's best interest with risks of DVT, possible increased risk of breast and endometrial CA, surgical risk
- GA may increase social stigma of person with disability by an unusual -childlike appearance
- GA may violate the person's identity and bodily integrity – failing to respect that person as they are and maintain bodily integrity
- GA may violate the universal right to dignity and respect (UN Convention on Rights of Persons with Disabilities 2006)
Ethical Arguments Against Growth Attenuation

- There are limits to Parental Autonomy
- Parent's personal interest to lessen the burden of caregiving may be motivation
- Parents may have an unrealistic view of the impact of GA on their lives
- Parents may just want control in the lives of their disabled child when they otherwise feel powerless


Growth Attenuation Therapy (GAT) for Girls with Severe Physical and Cognitive Disabilities (SPCD)

- Have you prescribed GAT for girl(s) with SPCD?
  - 1. YES
  - 2. NO
Growth Attenuation Therapy (GAT) for Girls with Severe Physical and Cognitive Disabilities (SPCD)

• Have you consulted an Ethics Committee about GAT for girl(s) with SPCD?
  • 1. YES
  • 2. NO

Ethical difference? Precocious Puberty

• Many children with profound developmental delay experience precocious puberty – is it ethically permissible to forgo intervention for these children with the intent to attenuate growth?
• Some would say a medical intervention such as GA should not be used to "move someone away" from the norm, that is to stunt growth
Growth Attenuation Therapy (GAT) for Girls with Severe Physical and Cognitive Disabilities (SPCD) and Precocious Puberty?

• Have you been asked about GAT for girl(s) with SPCD and Precocious Puberty?
  • 1. YES
  • 2. NO

Growth Attenuation Therapy (GAT) for Girls with Severe Physical and Cognitive Disabilities (SPCD) and Precocious Puberty

• Have you discussed foregoing interventions for girl(s) with SPCD and Precocious Puberty?
  • 1. YES
  • 2. NO
Growth Attenuation Therapy (GAT) for Girls with Severe Physical and Cognitive Disabilities (SPCD)

- Pediatric Endocrine Society (PES) 284/1100 member = response rate 26%
- 34% had been asked about GAT
- 34% had obtained ethics consultation for GAT
- 46% had withheld tx for Precocious Puberty if SPCD
- 11% had prescribed GAT for SPCD
- Calculated ~65 children with SPCD tx’d for GAT


Finding compromise – A middle ground

- Third parties feel very strongly on both sides of the debate – should we allow them to affect individual families or children?
- Case by case basis
- Allow an educated, informed decision between a caring set of parents in consultation with a competent physician with support from an experienced ethics committee
Clinicians’ Roles

• Providing information

• Help parents identify decisions that can be deferred (not possible in case of children with persistent, profound developmental and intellectual disabilities)

• Actively engage the parents in discussion

Clinicians’ Roles

• Engaging parents may mean many things:
  • Challenging the parents’ reasons
  • Sharing the provider’s own clinical and ethical opinions
  • Making recommendations
  • Exploring options that the provider might not deem optimal
  • Sometimes refusing parental requests
  • May need to refer to another clinician

Safeguards for appropriate use of Growth Attenuation

- Strict evaluation of developmental level and long term prognosis for improvement
- Consultation with Endocrinologist (and Gynecologist)
- Ethics consultation to determine if this treatment is right for the patient and family
- Guardian ad litem/Court involvement
- Consider doing under research protocol with oversight of IRB to gain data or at least with registry

Growth Attenuation Therapy (GAT) for Girls with Severe Physical and Cognitive Disabilities (SPCD) and Precocious Puberty

- Based on information presented at this NASPAG debate, would you be willing to discuss foregoing therapy for precocious puberty?
  - 1. YES
  - 2. NO
Growth Attenuation Therapy (GAT) for Girls with Severe Physical and Cognitive Disabilities (SPCD) and Precocious Puberty

- Based on information presented at this NASPAG debate, have you changed your opinion for or against GAT?
  - 1. YES
  - 2. NO

Safeguards for appropriate use of Growth Attenuation