The “Big Bang” Theory and Down Syndrome

Robert Perske

I am convinced that some of the most joyful, life-changing conventions in our field are those that focus on families with children having Down syndrome. Whole families come. Some appear with babes in arms. Clinicians named Pueschel, Crocker, Kumin, Falvey, Rosenberg and many others hold impromptu clinics in halls, lobbies, and special session rooms. It all goes so well because these parents hunger to learn everything they can about their own child with this disability.

There are sharing sessions galore for moms, dads, brothers, sisters, and grandparents. There are even sessions for persons with this disability who help one another to speak for themselves. Then come the banquets and the laughter and the awards and the cheering and the dances where all are dressed in their very best clothes. Many leave exhausted from these carnivals of hope, education, and comradeship, but they leave knowing that they are part of a powerful, expansive movement.

For me, speaking in sessions to these persons has always been a fun thing to do. It is a far cry from standing before some groups of well-résuméd, highly-certified professionals who often respond to all the points being made like cattle watching a passing train.

One can’t help but wonder if some earlier “big

(Growth Attenuation and Indirect-Benefit Rationale

Dick Sobsey

An article published by Gunther and Diekema in the October 2006 issue of Archives of Pediatrics and Adolescent Medicine reports on the use of high-dose estrogen therapy and hysterectomy for a six-year-old girl with severe and multiple disabilities. The stated purpose of the therapy is to stop the girl from growing, thereby keeping her weight down in order to reduce future lifting and transferring demands on her parents. The authors cite the American Academy of Pediatrics goal of eliminating institutional care of children by 2010 as a rationale for this treatment, suggesting that keeping the child small will increase the probability of her staying in her family home and allow for better parent-child interactions.

The primary benefit offered by growth attenuation is the potential to make caring for the child less burdensome and therefore more accessible.

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Growth Attenuation and Indirect-Benefit Rationale

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for the child less burdensome and therefore more accessible. A smaller person is not as difficult to move and transfer from place to place. Although this may seem to be an advantage that accrues to the caretakers rather than the child, it offers several distinct benefits to the child as well. (Gunther & Diekema, 2006, p. 1016).

According to the authors, “[b]ecause we have no direct experience with high-dose estrogen treatment in young children, the possible adverse effects and risks are difficult to assess with certainty” and the effects on “young prepubertal children will be significant” (Gunther & Diekema, 2006, p. 1015). Nevertheless, they argue that what is good for the family is good for the child.

An editorial commentary published in same issue (Brosco & Feudtner, 2006) raises a number of ethical questions and concludes that the procedure is “ill-advised” (p. 1078) for a number of important reasons. Nevertheless, this case report raises a more general question: Can medical and surgical interventions with significant risk to the individual with intellectual disabilities, ever be justified by indirect benefits to the individual when most direct benefits accrue to other caregivers such as family members?

With growth attenuation the presumed relationship between intervention is particularly complex and involves four steps: (1) medical and surgical intervention is predicted to attenuate height, (2) attenuated height is assumed to result in attenuated weight, (3) attenuated weight is assumed to reduce lifting and transferring demand on parents, and (4) reduced demands are expected to result in better parenting. Each of these steps adds complexity and adds additional risk of failure to achieve the stated goal.

There are many other examples of the use of interventions to achieve indirect benefits.

One fairly common example is when families members seek medications for individuals with intellectual disabilities to reduce hyperactivity or regulate sleep patterns...to reduce caregiving demands or allow time for their own sleep.

This example raises three important questions: (1) Is the effect of the medication normalizing? Using medication to increase the child’s sleep period from 6 hours to 8 hours per day may be easier to justify than increasing it from 8 hours to 12 hours per day. Reducing hyperactivity to a level that allows greater participation in normal activities provides some direct benefit to the individual, reducing activity to a point that makes participation more difficult is probably harmful. (2) What is the potential harm to the individual? In addition to the obvious risks and side effects of medications this may include social impacts. (3) Are the proposed indirect benefits likely to be realized? The massive overmedication and incapacitation of people with developmental disabilities in institutions should serve as a grim reminder that medicating people with disabilities can enable neglect as well as better care.

In a more extreme example of indirect benefit argument, in some parts of the world parents or other caregivers have requested that physicians perform amputations or medical mutilations for children with disabilities in order to make their children more productive beggars (e.g., Sharma, 2005). Many of these families suffer from extreme poverty and raising a child with a disability adds to the financial woes of the family. Children with severe disabilities are unlikely to be employable or marriageable as adults. Making the child a more productive beggar adds family income and this may allow them to continue caring for the child for a longer period of time. For families facing extreme poverty, this motivation to improve income may be difficult to resist. While the prospect of mutilating a child is horrific, the alternatives for

We should consider whether the presumed indirect benefits of better and more enduring care are likely to refult from the intervention...

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Transplants for Developmentally Delayed Children

David Magnus, Holly Tabor, and Katrina Karkazis

Stanford University and the Lucille Packard Children’s Hospital (LPCH) hosted a gathering of leading figures in pediatric and transplant ethics to address the difficult challenge posed by transplant listing decisions for developmentally delayed children. The goal of the gathering was to produce a consensus statement that will serve as guidance for listing teams.

The idea for holding the meeting came out of a number of difficult ethics consults that took place at LPCH. The liver team in particular hoped for the LPCH ethics committee to provide guidance on listing decisions. A subcommittee was formed, and the group quickly realized there was little data on how such decisions are made in practice. An initial survey of transplant programs was conducted prior to the meeting and the data was shared with the group. It showed that different programs de facto have differing standards, that many programs routinely deny transplants to patients who are even moderately delayed, and that there are some cases where children are denied access to transplant because of their mental status even though they are only mildly delayed.

The meeting began with a presentation by Dr. William Bronston, from the National Working Group on Disability and Transplantation. Bronston has been a tireless advocate on behalf of disabled patients seeking access to transplants and expressed the challenge many parents face in trying to obtain needed medical services for their children. Chris Richards, a Stanford medical student who has collected the data on transplant listing decisions at pediatric institutions provided the preliminary data. The data included several hypothetical scenarios including one with a mildly retarded child who had no relevant medical contraindications and who needed a transplant (slightly different versions of the scenario were developed for kidney, liver, and heart transplant centers). The results of the survey clearly demonstrated that even mildly impaired children (e.g., an adolescent with a second grade reading level) would be denied access to transplant by some services, particularly for heart transplant. Finally, Karen Wayman, PhD, from the LPCH liver team explained how to do a developmental assessment and worked through several examples with the group.

Many felt that it is fundamentally unjust to take cognitive status into account in listing decisions.

While the group seemed to reach agreement that it was highly problematic that children who are mildly to moderately delayed are denied access to transplant (which seems not uncommon for at least some organ systems), the group was unable to reach consensus on whether there (Continued on page 4)
was a point where there is sufficiently little benefit to the patient to justify transplantation. Most of the group seemed to feel listing patients in a permanently unconscious state would be inappropriate, but beyond that there was no consensus. However, there was agreement about a number of process issues that would ensure that each individual patient receives a full and fair assessment to determine the value of the transplant for them (and from the patient’s perspective). The problem of unfair discrimination that seems on its surface to be taking place may best be addressed by improvements in the processes that teams use in listing decisions.

The group is in the process of producing a set of recommendations about how teams should address developmental delay and will be publishing the results. In addition, Chris Richards and the group at Stanford is collecting more data and will be publishing the results of their surveys as well as follow-up, in-depth interviews that will be conducted.

The gathering, supported by a grant from the Greenwall Foundation (David Magnus, PhD, Principal Investigator), included representatives from Stanford and LPCH transplant surgery, ICU, nephrology, cardiology, and the liver teams; members of the LPCH ethics committee and a parent of a developmentally impaired child. Attendees included Alex Kon, MD, from UC Davis; Art Caplan, PhD, from the University of Pennsylvania; Chris Feudtner, MD, PhD, from Children’s Hospital of Philadelphia and UPenn; Doug Diekema, MD, from the University of Washington and Seattle Children’s Hospital; Rick Kodish, MD, from the Cleveland Clinic; Joel Frader, MD, from Northwestern; Lainie Friedman-Ross, MD, PhD, University of Chicago; Mark Fox, MD, University of Oklahoma; Robert “Skip” Nelson, MD, PhD, CHOP and UPenn; Robert Truog, MD, Harvard and Boston Children’s Hospital; Stuart Youngner, MD, Case Western Reserve; Robert Veatch, PhD, Georgetown University; Ben Wilfond, MD, the University of Washington and Seattle Children’s Hospital; and Larry Nelson, JD, PhD from Santa Clara University. The LPCH ethics committee co-chairs, Julie Collier, PhD, and Christy Sandborg, MD, chaired the meeting.

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bangs” in history gave rise to such an expansive movement. I say yes. I can pinpoint at least two such explosions. These explosions were heart-ripping. Even so, the way the folks in this field gathered around these wounds and worked for healing was fantastic.

Washington, DC

The John F. Kennedy Center for the Performing Arts, October 16, 1971

People from all over the world arrived for the International Symposium on Human Rights, Retardation and Research. The Eisenhower Theater was packed with persons eagerly waiting for the opening plenary to begin. When it did, a film was shown that shook many in the audience right down to their socks.

The film was entitled, “Who Should Survive?” It focused on a real newborn baby boy in the maternity ward of Johns-Hopkins hospital. The child was born with Down syndrome and an intestinal block. The latter could be corrected with a fairly simple operation. So the physician asked the parents for permission to operate. The latter could be corrected with a fairly simple operation. The parents refused. They said that it would be unfair to their other two so-called normal children to be brought up with a “mongoloid” sibling. The infant was moved to a corner in the hospital nursery and a sign was hung on the crib that said, “Nothing by Mouth.”

From time to time the father telephoned to ask, “How are things going?” The film showed the anguish on the faces of the caring staff. The senior pediatric resident in charge of the infant’s care stated, “I tried not to look
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at the baby and when I did, I didn’t want to touch it.” Later, he said, “It took 15 days for the baby to become dehydrated enough to die. That was an awfully long time.”

After the film, six famous experts representing key legal, moral, ethical, social, public policy and mental retardation perspectives gathered in a circle on the stage. They faced each other and discussed the film. At first, the circle widened as each expert tried to define the situation in his or her own professional language. Then they drew closer, trying to speak a common human language about a neglected, dying baby. When the closeness became too painful, one of them would “blow it” by making an asinine remark. For example, during one moment of such closeness, a physician blurted out, “What about the war in Vietnam?” The audience clapped during one moment of such closeness, a physician blurted out, “What about the war in Vietnam?” The audience clapped. The request was granted.

Many times those who become the greatest among us, do so out of personal needs to compensate for limits in their lives.

So, under the wings of The Arc, the National Down Syndrome Congress began to take shape. It gathered strength and became more alert in case another tragedy like the death at Johns-Hopkins took place. Sure enough, it came.

**Bloomington, Indiana, Bloomington Hospital, April 9, 1982**

“Infant Doe,” a baby boy with Down syndrome was born with an esophageal atresia, the separation of the esophagus from the stomach. Again, doing the life-saving surgery was no problem. Again, parents refused to permit the operation. Again, an infant was shoved into a corner and starved to death.

This time, however, the funk surrounding the issue was less pronounced. Numerous families contacted the hospital begging for permission to take the baby and raise it as their own. Persons having children with the syndrome made heart-warming statements about them. National syndicated columnist George Will spoke out on the issue:

Jonathan Will, 10, fourth grader and Orioles fan (and the best Wiffle-ball hitter in Southern Maryland), has Down syndrome. He does not “suffer from” Down syndrome. He suffers from nothing, except anxiety about the Orioles’ lousy start. He is doing nicely, thank you. But he is bound to have quite enough problems dealing with society – receiving rights, let alone empathy. He can do without people like Infant Doe’s parents, and courts like Indiana’s asserting by their actions the principle that people like him are less than fully human. On the evidence, Down syndrome citizens have little to learn about being human from the people responsible for the death of Infant Doe.

This time, movements on behalf of persons with Down syndrome “pulled out the stops.” They filled the media with positive letters to the editors. They approached legislators. They reached the ears of President Ronald Reagan who was shaken by the news. He discussed the issue in depth with Surgeon General C. Everett Koop. Since then, a cascade of legislative acts and judicial judgments has protected citizens with Down syndrome as never before.

I also experienced shock waves coming from this perplexing issue. In response, I spent time talking to families with children having Down syndrome. I even stayed overnight with some. Consequently, I wrote the novel, *Show Me No Mercy*, with all of the characters in it being fashioned after real people. Even so, the solution to the Bloomington explosion came long before the book came off the presses.

**American Journal of Obstetrics and Gynecology, March 2005**

A study in this journal may fuel another explosion regarding the life and death of infants with Down syndrome. Or it may not. The Harvard University Gazette billed the study as “the largest and most comprehensive study on prenatally diagnosed Down syndrome to date.” It “estimates” an abortion rate of 80 to 90 percent when prenatal screening reveals the possibility for the condition.

Interestingly, there are now three national agencies standing ready to respond – National Down Syndrome Congress, National Association on Down Syndrome and National Down Syndrome Society. I don’t know how this came to be, but I do know that

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many parents and professionals hold memberships in all three. From them a chorus of early responses can now be heard:

- Most expectant mothers only learn about the painful side of such a birth from clinicians.
- Many admitted that they felt pressure from physicians to terminate the birth.
- Almost never did they receive the latest information on Down syndrome.
- Almost never did they receive information about organizations for persons with Down syndrome.
- Prenatal Down syndrome testing is wrong 20 to 40 percent of the time.
- Most young expectant mothers carry such hope and joy regarding the baby within them, they refuse all offers of prenatal testing.
- One organization claims that these children bring more love in the world.

Of course obstetricians are more prone to talk about “perfect births.” We wouldn’t go to one who did not possess this high goal.

Actually, everybody longs for Superbaby, but nobody gets one. Many times those who become the greatest among us, do so out of personal needs to compensate for limits in their lives. And their success is often an utter surprise to their parents – mothers and fathers who felt that this one was too sickly, slow, nervous, loud or strange.

Robert Perske is an advocate and the author of Circle of Friends, Deadly Innocence and Unequal Justice.

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Words for a Child With Down Syndrome

(Written on the sleepless night after watching the documentary, “Who Should Survive?” Washington, DC, October 16, 1971)

Robert Perske

We didn’t want you, Helpless child of fifteen days
You were in the way, So we shoved you into a corner ... And let you starve to death.

No one wanted you, Helpless child of fifteen days. Not your parents Nor those who speak for God, Nor the “helping” professionals Nor men using their wits to uphold justice.

Why didn’t we want you Helpless child of fifteen days?

Would you have stood in the way of our mad chase After the ideal family? (Which none of us will ever achieve.)

Would we trip over you as we raise higher The GNP as our Holy Grail? (We must be “Number One” you know.)

Is it because we have never learned To live graciously with our failures? (Failure is such a hard thing to face.)

Is it because your less-than-perfect presence Reminds us of our drives to “be something.” (We never want to be “a nothing.”)

You bothered us, Helpless child of fifteen days.

We had to reject you Because you made too obvious something in ourselves ... That we just didn’t want to face.
these families may be tethering, leaving the child alone at home tethered to a bed while both parents go out to work for eight to twelve hours each day.

Of course, these justifications should not be interpreted as an endorsement for mutilating children, they are merely intended to suggest that the same kind of indirect-benefit arguments that support growth attenuation could be applied to a very wide range of interventions such as amputation or mutilation. The most compelling question raised by the mutilation issues may be “why is there no humane and normalizing alternative available?” We might ask the same question about growth attenuation.

While the hysterectomy and high-dose estrogen does not result in the immediate disfigurement associated with amputation, the long-term effects of high dose-estrogen applied to a six-year-old child are likely to result in highly atypical physical appearance that is at least as dramatic as simple amputation. The effects are likely to include extremely short stature, infantilization of long-bone body proportions, breast enlargement, acne, and, ironically, increased body fat and weight gain.

Although Gunther and Diekema’s stated purpose of growth attenuation is to minimize weight, high-dose estrogen has frequently been associated with weight gain. While the effects of estrogen on body weight and accumulation of body fat are complex, high-dose estrogen often results in weight gain. For example, Atarés, Zachman and Prader (1986) used estrogen to stop 36 tall adolescent girls from growing taller. They also recorded the effect on the girls’ weight and compared it to the expected gain based on the girls’ previous growth. Although the treatment successfully attenuated their growth in stature, it actually accelerated weight gain, which doubled compared to their projected weights without treatment. The average weight gain in the first 12 months of treatment was 19.8 ± 7.9 pounds (Atares, Zachman, & Prader, 1986). The longer the treatment was continued, the greater the weight gain.

Of course, the weight-gain effects of estrogen might be curtailed by severe restriction of food intake. It would likely need to be severe since estrogen has a specific effect of adding adipose tissue in specific areas of the body, and the data suggesting that high-dose estrogen doubles weight gain suggests that greater food restriction would be required after estrogen treatment than would be required without it. If actual weight gain reduction is going to be accomplished through severely restricted calorie intake, why subject the child to risks of drugs and surgery?

We should consider whether the presumed indirect benefits of better and more enduring care are likely to result from the intervention, assuming that the treatment actually achieves weight control. Lifting and transferring older children and adults with disabilities does place a real demand on caregivers and that demand increases with the weight of the individual who must be lifted or transferred (e.g., Brown, Mulley, 1997), but mechanical lifts, caregiver training, and exercise have been shown to reduce demands and injuries to caregivers, and these may be better alternatives that attempting to limit height and weight of children. In addition, while the demand of lifting is one factor in caregiver stress, it is only one of a large constellation of factors (e.g., financial impact, child behavior, social supports, access to educational programs) that affects caregivers and there is no evidence that lifting is a critical factor in the quality of interaction of parents and their children with disabilities or in out of home placements. In addition, research suggests that caregivers’ perceptions or appraisals of the demands they face are a much better predictor of perceived burden than objectively measured demands or stressors (e.g., Miltiades, & Pruchno, 2001). In short, there is no reason to believe that a child’s weight is a critical variable in quality of care or in probability of out-of-home placement.

Considering that growth attenuation and hysterectomy have substantial health risks, are unlikely to produce significant attenuation of weight gain, will probably result in atypical physical appearance, and may actually lead to increased weight, it is likely to do harm to the child and unlikely to achieve much of its stated benefit to the parents. Considering that there is no direct evidence to suggest that controlling a child’s weight is a significant factor in reducing the probability of institutionalization or in increasing the quality of care, there is no evidence-based reason to believe that the assumed direct benefit to the parents, if achieved, would translate into the proposed indirect benefit to the child.

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The indirect-benefit ethical rationale for growth attenuation, like the indirect-benefit arguments presented for amputation and mutilation or sedation, should be carefully scrutinized. While there are undoubtedly legitimate circumstances in which indirect benefits can justify medical interventions, the rationale should always be approached with the greatest of caution, particularly when the supposed indirect beneficiary is a child with a disability or other vulnerable person.

References


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Bibliography

Harriet Hutson Gray

These books and articles have been added recently to the collections of the National Reference Center for Bioethics Literature (NRCBL). Similar citations may be found online by searching PubMed database at the National Library of Medicine and the ETHX on the Web database at the NRCBL. Access and tips for searching may be found at the NRCBL website at http://bioethics.georgetown.edu.


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