Transition from Pediatric to Adult Epilepsy Care: A Difficult Process Marked by Medical and Social Crisis

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Disclosure

Nothing to disclose.
What do you need to be convinced of?

• Epilepsy can mess up normal adolescence.
• Children with epilepsy often have social problems as adults even if their epilepsy remits.
• Transition to adult care takes time and energy and no one knows how to do it best.
Early childhood  Adolescence  Adulthood

Strong association with mental handicap and neurological deficit

Especially JME and symptomatic partial
It is a long from childhood to old age
Erik Erikson

Adolescence
Young Adulthood
Adulthood
Adolescence – identity vs identity confusion
FIDELITY

SEX, DRUGS and ROCK and ROLL
Young Adulthood – intimacy vs isolation

LOVE
Adulthood – generativity vs stagnation
CARE
Why might epilepsy disrupt adolescence and mess up adult life?

- **Autonomy** – becoming independent
- **Body image** – a big, big concern
- **Peer group** – what could be more important?
- **Identity** – who am I?
Autonomy

- Think of the things that I can no longer do without supervision – take a bath, swim, stay up late, drink beer, drive, have babies, become a pilot, join the armed forces, take aspirin, scuba dive, go to dances with strobe lights, play video games, go on the school trip.
- Think of all the things that I have to do – everyday, yes everyday - take my pills twice a day, go to the stupid doctor, have EEGs that mess up my hair, watch my weight.
There is something terribly wrong with me (remember I have never seen my seizures).

The pills will somehow mess up my hormones.

The pills will somehow mess up my bones.

The pills will make me fat.

My hair will fall out.
Peer Group

- No will go on a date with me
- No one else has this stupid problem
- No one else has the tremor from VPA
- How can I avoid the strobe lights and ever go out in public?
- What happens if I have a seizure at school and wet my pants?
- No one will ever marry me
Identity

• I am not supposed to do most of the things that my friends do – drinking, drugs, sex (the pill apparently does not work)
• There are many professions that I am blocked from – how will I cope with life?
• I will die during sleep from SUDEP
Are these concerns realistic?

2001 US survey: 19,441 high school students

Austin *Epi Behav* 2002:3;368

- 19% - epilepsy is a mental illness, 30% unsure
- 50% - unsure if PWE could drive cars, marry, have children or work
- 51% - people die during seizures
- 33% - thought they knew what to do if someone had a seizure
- 50% - unsure if epilepsy is contagious
- 33% would date someone with epilepsy
- 50% - you can look at a person and tell if they have epilepsy
Fear of People with Epilepsy

Jane accidentally bumped into John, who has epilepsy, in the cafeteria. John became very angry and stabbed Jane repeatedly with a fork. He explained that he was not to be blamed because he was suffering from an epilepsy seizure. Is this believable?

– Very likely
– Possible
– Unlikely
• Medical and law students, physicians, childcare workers, general public and people with epilepsy ($n=271$ in 1981 and $n=388$ in 2006).
• Nearly half of people believed that violence was possible or likely during a seizure.
• All groups answered at least 40% of questions incorrectly, except for physicians who answered 20% incorrectly.
• Responses were fairly stable over the 25-year interval.

Collins Epi Behav 2007;10:69-76
ANATOMY OF A TEENAGER'S BRAIN

THE BIRDS AND THE BEES LOBE

REBELLION CENTER

SUPER TURBO REBELLION CENTER

SELF IMAGE

FITTING IN GLAND

CENTER OF UNIVERSE CENTER

EVERY EPISODE OF THE SIMPSONS

INDESTRUCTIBILITY CORTEX

MEMORY FOR MUSIC

MEMORY FOR CHORES, HOMEWORK, ETC.

LOVE FOR PARENTS

SLANDER PREREX

CAR KEY'S CRINGING

ABILITY TO BE SEEN IN PUBLIC WITH PARENTS

PEER PRESSURE RESISTANCE

PLANE TO BRUSING

Judgement Gland

Slang Decoder

"Cool" Gauge

Memory for Parents

Slang for Parents

Greed for Answers

Slang for Answers

Slang for Parents

Greed for Answers
What constitutes a good social outcome for someone with normal intelligence?

Jalava, Sillanpaa *Epilepsia* 1997; 38:708

- **Employment** with financial self-sufficiency
- **Married** or cohabiting
- Participation in **social life**
- One or more **hobbies/activities** outside home
Prevalence cohort, 35 yrs follow up. 100: “epilepsy only”

C/W random sample of Finnish adults matched for sex, age and place of birth, “epilepsy only” had lower education, employability and marriage rate.

C/W an occupational control group, “epilepsy only” rated their own ability to control their lives as “poor or missing” four times more often.

Polytherapy and continuing seizures were associated with reports of decreased life satisfaction and poor general health.
What happens to children with Childhood Absence Epilepsy when they become young adults?

- Follow up all with CAE to age ≥18 yrs.
- Compared with Juvenile Rheumatoid Arthritis (JRA) as a chronic disease control.
- Structured interview.

Wirrell Arch Pediatr Adolesc Med 1997;51:152
Follow up to >18 years of age

<table>
<thead>
<tr>
<th></th>
<th>Absence n=56</th>
<th>JRA n=61</th>
</tr>
</thead>
<tbody>
<tr>
<td>Female</td>
<td>68%</td>
<td>62%</td>
</tr>
<tr>
<td>Mean age onset</td>
<td>7.4 yrs</td>
<td>6.7 yrs</td>
</tr>
<tr>
<td>Age f/u</td>
<td>23 yrs</td>
<td>23 yrs</td>
</tr>
<tr>
<td>Remission</td>
<td>57%</td>
<td>28%</td>
</tr>
<tr>
<td>Still on medication</td>
<td>32%</td>
<td>38%</td>
</tr>
<tr>
<td></td>
<td>Absence</td>
<td>JRA</td>
</tr>
<tr>
<td>-------------------------------</td>
<td>----------</td>
<td>----------</td>
</tr>
<tr>
<td></td>
<td>n=56</td>
<td>n=61</td>
</tr>
<tr>
<td>No high school grad</td>
<td>36%</td>
<td>14%</td>
</tr>
<tr>
<td>Behavior</td>
<td>41%</td>
<td>10%</td>
</tr>
<tr>
<td>Unplanned pregnancy</td>
<td>34%</td>
<td>3%</td>
</tr>
<tr>
<td>Heavy drinking</td>
<td>39%</td>
<td>16%</td>
</tr>
<tr>
<td>Psychiatric</td>
<td>54%</td>
<td>31%</td>
</tr>
<tr>
<td>Unskilled labor</td>
<td>53%</td>
<td>16%</td>
</tr>
</tbody>
</table>
Absence vs JRA

• In statistical modeling epilepsy remission accounted for small amount of the adverse outcomes; however those with ongoing seizures had the worst outcome.
• Normally intelligent young adults with CAE have a lot of serious social problems whether or not their epilepsy is outgrown.
Other IGE syndromes from the Partial and Convulsive Group

- Juvenile Myoclonic Epilepsy – **JME n= 24**
- Idiopathic Generalized Epilepsy with Generalized Tonic-Clonic Seizures Only – **IGE – GTC n=40**
- Follow up for 20-35 years

Camfield Neurol ogy 2009;73:1041
Camfield Epi Behav 2010;8:61
<table>
<thead>
<tr>
<th></th>
<th>IGE GTC</th>
<th>JME</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Typical Age of onset</strong></td>
<td>4-15</td>
<td>10-20</td>
</tr>
<tr>
<td><strong>Neurological and</strong></td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td><strong>intellectual examination</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>GTC</strong></td>
<td>√</td>
<td>√</td>
</tr>
<tr>
<td><strong>Absence</strong></td>
<td></td>
<td>√</td>
</tr>
<tr>
<td><strong>Myoclonus</strong></td>
<td></td>
<td>√</td>
</tr>
<tr>
<td><strong>Imaging</strong></td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td><strong>EEG spike discharge</strong></td>
<td>Gen S-W &gt;2.5 hz</td>
<td>Gen S-W usually fast</td>
</tr>
<tr>
<td><strong>Photosensitivity</strong></td>
<td>Uncommon</td>
<td>1/3</td>
</tr>
</tbody>
</table>
### Seizure Severity

<table>
<thead>
<tr>
<th></th>
<th>IGE - GTC</th>
<th>JME</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Episodes of status</td>
<td>8%</td>
<td>38%</td>
<td>0.01</td>
</tr>
<tr>
<td>&lt;11 seizures</td>
<td>53%</td>
<td>26%</td>
<td>0.03</td>
</tr>
<tr>
<td>&gt;21 GTC</td>
<td>17%</td>
<td>61%</td>
<td>0.0005</td>
</tr>
<tr>
<td>Remission (Off AEDs)</td>
<td>75%</td>
<td>38%</td>
<td>0.005</td>
</tr>
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</table>
### Social Outcome

<table>
<thead>
<tr>
<th></th>
<th>IGE-GTC</th>
<th>JME</th>
</tr>
</thead>
<tbody>
<tr>
<td>High School Grad</td>
<td>60%</td>
<td>87%</td>
</tr>
<tr>
<td>Unemployed</td>
<td>33%</td>
<td>31%</td>
</tr>
<tr>
<td>Depression (drug)</td>
<td>20%</td>
<td>39%</td>
</tr>
<tr>
<td>Living alone</td>
<td>23%</td>
<td>30%</td>
</tr>
<tr>
<td>Pregnancy outside stable relationship</td>
<td>11/13 (85%)</td>
<td>11/15 (73%)</td>
</tr>
</tbody>
</table>

P = 0.002
## Social Outcome

<table>
<thead>
<tr>
<th></th>
<th>IGE – GTC</th>
<th>JME</th>
</tr>
</thead>
<tbody>
<tr>
<td>≥ 1 adverse social outcome</td>
<td>76%</td>
<td>76%</td>
</tr>
</tbody>
</table>

**Adverse social outcomes** = no high school graduation, unemployment, psychiatric diagnosis, no romantic relationships, living alone, conviction of a crime, pregnancy outside of a stable relationship
Symptomatic Generalized Epilepsy – SGE
12% of all childhood onset epilepsy

- Many have mental handicap
- What is a good social outcome for someone with mental handicap?
What constitutes a good social outcome for someone with mental handicap? (Miller, Chan J intellect Disabil Res 2008;52:1039)

- **Social support** – large network consisting of socially supportive staff and family and friends
- **Self-determination**/perceived control
- **Work/productivity** (competitive and supported employment is better than sheltered workshop or home).
- **Adaptive behavior** – independent living skills, social skills, communication + academic skills
Most common SGE syndromes

- West
- Lennox Gastaut
- Myoclonic/astatic
- Dravet
- Undefined
SGE 20 years after diagnosis

- Dead: 25%
- Remission off AEDs: 25%
- Remission on AEDs: 6%
- Continuing seizures with AEDs: 8%
- Intractable: 36%

Camfield Dev Med Child Neurol 2008;50: 859
SGE social outcome if ≥18 yrs at end of follow-up: n=45

**Good** 18% n= 8: judged able to live independently

**Fair** 22% n=10: cannot live independently but do not require total care

**Poor** 60% n=27: require assistance with all activities of daily living

- Only 3 young adult survivors with SGE were intellectually and neurologically normal, seizure-free/off AEDs, financially independent and living on their own with a partner.
SGE: assessment by adult neurology – not a criticism, just an observation

- Canada – no financial barriers to access.
- 75% of survivors had never seen an adult neurologist.
- 18/25 (72%) with intractable epilepsy had never seen an adult neurologist and 52% had never received a “newer” AED after VPA.
How can children with epilepsy find their way to a more successful adulthood?
Important Distinction

- **Transition** – a process beginning in childhood to prepare youth with chronic illness and families for “adult” health care.

- **Transfer** – the formal handing over of care from a pediatric to adult health care system.
Transition – problems

• Differing cultures
  Adult medical care focuses on the **individual**. Pediatric medical care is very **family-centered**.
• Families are reluctant to move on to adult care.
• Pediatric specialists have trouble saying goodbye and resist passing along patient care to a system perceived as less proficient.
• Difficulty finding knowledgeable adult primary care providers and subspecialists.
• Lack of institutional support, time and reimbursement.
Literature review + interviews with 50 pediatric and adult epileptologists in 11 major centers in 7 countries.

- Little published peer-reviewed research (lots of editorials, no data).
- No center was satisfied with their transition/transfer process for children with epilepsy.
- **Perceptions**: Pediatrics - adult services have little interest in the co-morbidities that accompany childhood onset epilepsy. Adult Neuro - pediatric services are coddling and indulgent.
- No centre had data on the success of their program.
- 10/11 centers had a transfer process with no formal transition program.
Models for transition/transfer

No transfer

Continued follow-up in a pediatric setting.

• The pediatric specialist ages with the patient – puts off transfer until the specialist retires unless the institution is committed to a “life to death” program.
• Fosters dependency - everyone deserves to grow up
• Logistical problems for medical and social services – what happens if you have a complex epilepsy disorder and you need your gall bladder out at age 30 in an adult hospital and have seizures post op? No one there knows about your epilepsy problem.
• Lack of knowledge about adult diseases – hypertension, type 2 diabetes, COPD
Models for transition/transfer

- **Abandonment** (fend for yourself). The family physician takes full responsibility – a physician who has had little/no role during the pediatric years. The youth and family are not attached and doubtful of this physician’s competence. Few are epilepsy experts.

- **Adult rehabilitation program** These are oriented towards consultation and short-term intervention. Care is multidisciplinary but long-term follow up is unusual.
Models for transfer

• **Adult neurologist or adult neurology clinic.** (a consultation model with few follow up visits unless re-referred by family physician). Only a few special adult neurologists show interest in long term care for developmental brain disorders. One paper describes a Pediatric – Adult Nurse transition clinic before moving to the adult neurologist *(Jurasek J Neurosci Nurs. 2010;42:181.)*

• **Transition clinic** attended jointly by adult and child neurologists in the adult setting. Families like this idea. Without any objective data, *we think this is the best model*. It allows family comfort and dialogue between pediatric and adult neurology over several visits.
Managing the teenager with epilepsy: paediatric to adult care. Appleton *Seizure* 1997;6:27

- Description of 120 “transitioned” patients
- Clinic staff – pediatric neurologist, adult neurologist, nurse practitioner
- 37% had mental handicap and 45% had JME
- Most seen for 2 visits
- 10% did not have epilepsy, 22% had a change in medication
- See also Smith *Eur J Neurol.* 2002;9:373
Solutions to consider

**Transition:** Build skills that last for children and for parents. They should know enough about their epilepsy to know when to seek medical care and if the medical care is adequate.
Solutions to consider

- Find a willing adult service; start a transition program.
- Learn the realities/practice style of adult/pediatric neurology – spend a few afternoons in each others office or clinic.
- Talk with patients and families often about long-term health issues. Emphasize education, sexuality.
- Foster research to know what happens to pediatric epilepsy syndromes in adulthood.
- Encourage Epilepsy Support Groups to inform patients/families about new developments – can you be sure that anyone else do this?
Solutions to consider

• For the mentally handicapped
  - Guardianship/Trust Fund/Will
  - Involvement of many family members and friends who cross generations and can insist on comprehensive care and prevent abuse by demanding accountability.
    (avoid becoming “known well by no-one”)
Continue to be aware of the interaction between epilepsy and social development.

Adolescence

Young Adulthood

Adulthood