

Psychosocial Impact of Usher Syndrome

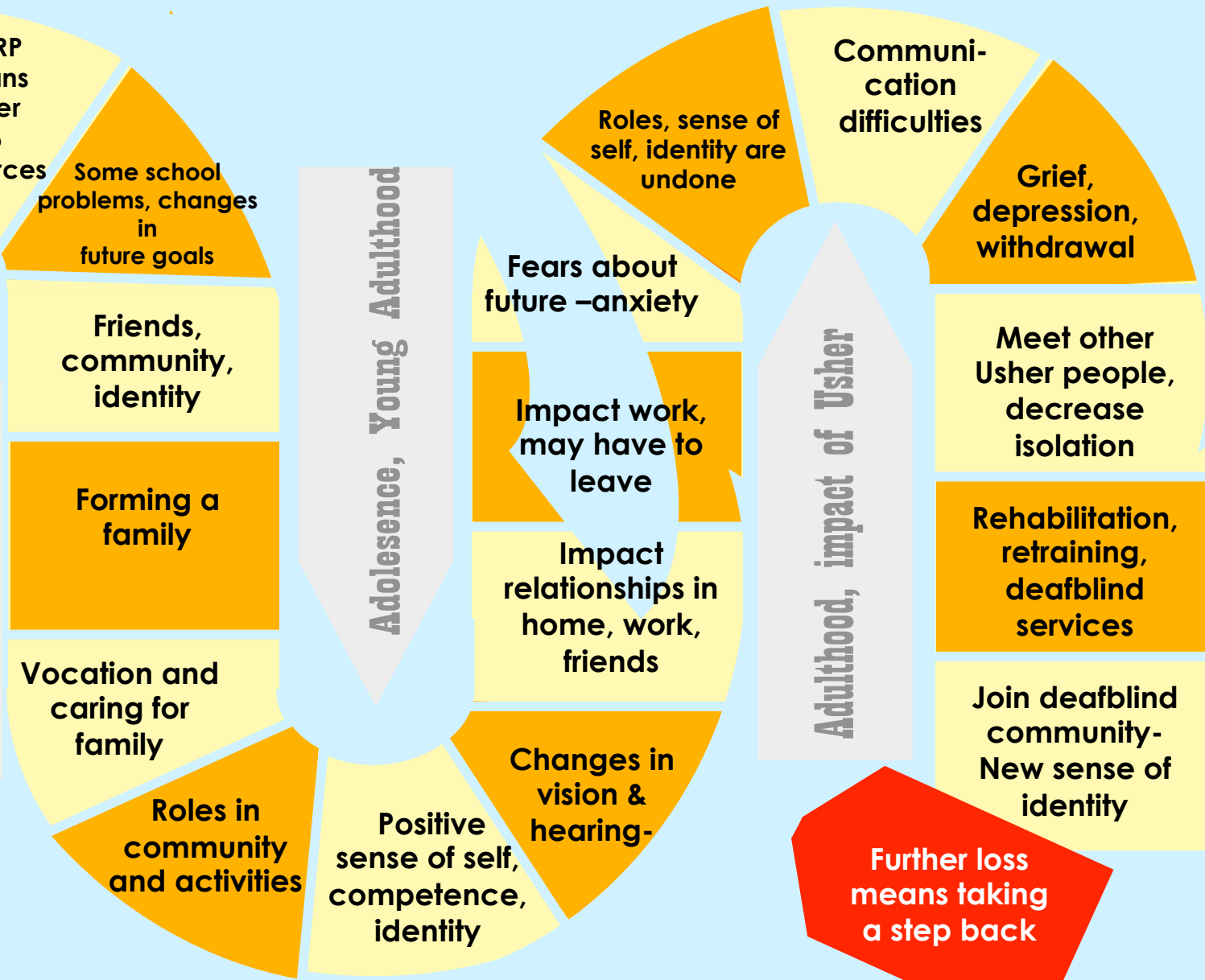
Adults and the Family

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Background

- In 2007, with the Information Center for Acquired Deafblindness, interviewed 13 people in Denmark, 10 with Usher and 12 people with Usher in the US about their lives, experiences, impact of RP, sense of identity.

ADOLESCENCE



Communication

Tactile
Sign
Language

Usher issues:
Communication,
Isolation, Loss,
Lack of information



Identity

- I am deafblind-
Seattle, WA – US I
- I am deafblind- and I am also deaf- US I
Copenhagen
- I am deafblind-
Denmark- US II
- I am deaf and visually impaired- US I
New York City
- I am hard of hearing and visually impaired
New York City- US III

Identity- Usher I

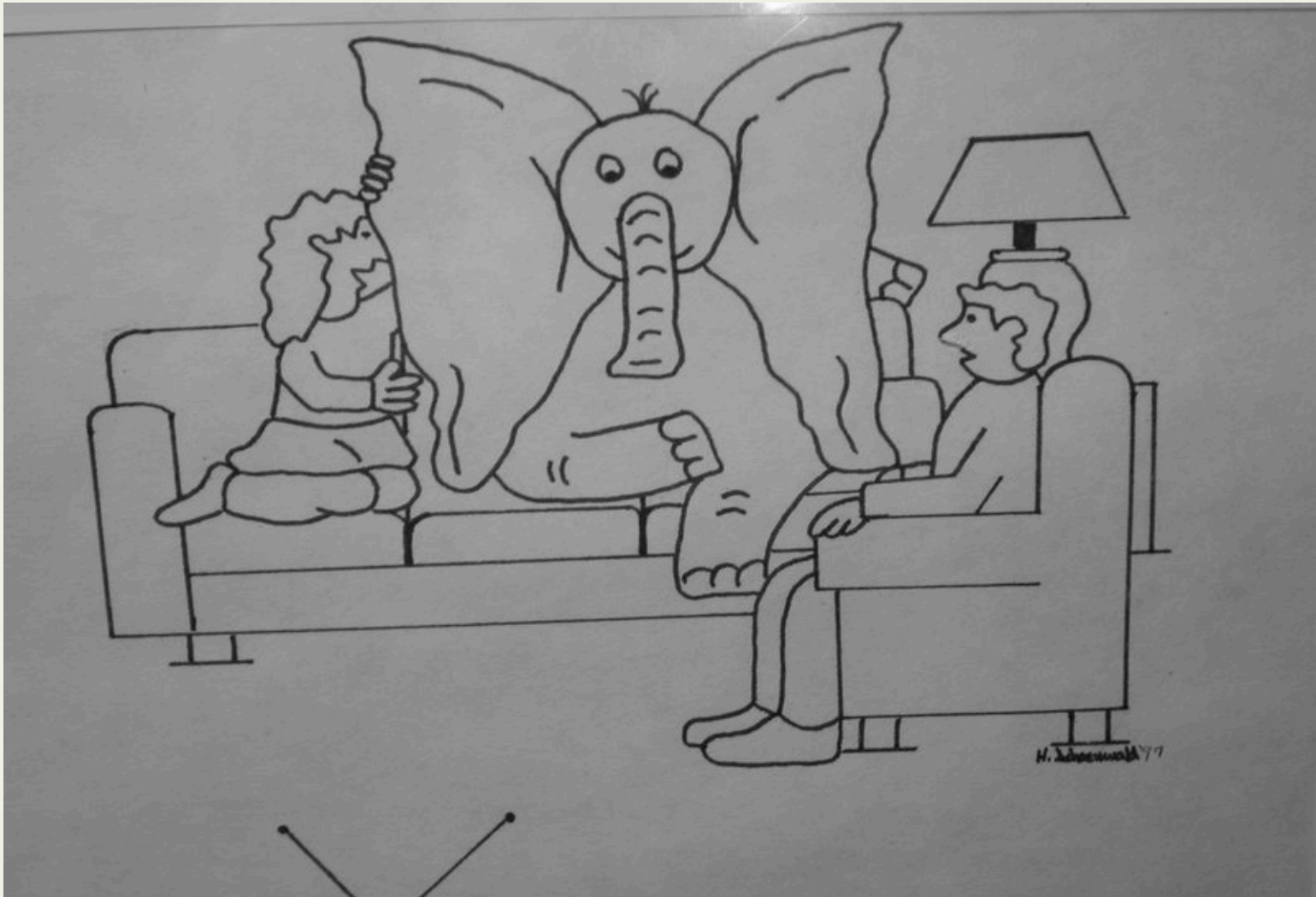
- Sign language, Deaf community, marry or partner Deaf
- May be changing due to cochlear implant
- Family of origin may not know sign language. Group identification different from parents
- Roles, activities, pastimes, supports from Deaf community
- Deafness is not disability- RP is the disability
- May know others with Usher I, but not Usher II

Identity Usher II & III

- Uses mainstream spoken language
- Identifies with mainstream culture
- Often marries or partners with hearing,
- Has compensated for hearing loss
- Often does not know any one with Usher
- Hearing loss and RP are both disabilities

The elephant in the living room

(drawn by Heather Schoenwald, Usher II)



Diagnosis of RP

- Already having problems
- Diagnosis is devastating
- Gone is the expected life, facing the unknown
- Finding information about living with RP is difficult
- Fears, anxieties, depression, suicidal ideation
- Reactions in Denmark and US were very similar

Impact on all spheres of life

- Sense of self, identity, competence
- Communication
- Vocational world
- Family Relationships
- Community and social relationship,
- Loss of activities, hobbies, pastimes
- Life becomes busy with professionals of all kinds

Family relationships

- Breadwinner and caretaker roles threatened
- Children avoid deafblind parent
- Deafblind person can be marginalized
- Sighted family member takes over roles
- Deafblind member grieves loss of roles, function, place in family
- Family activities become labored, require planning
- Some relationships do end
- Professionals So many of them

Communication

A Danish deafblind woman: “It’s hard to be deafblind; it can be exhausting because you have to use so many resources just to communicate, and hearing people just don’t understand.”

Tactile Sign needs to be modified and the pace is slower.

Equipment is necessary- FM systems, real-time captioning, speechreading is harder

The process can be exhausting

Community & social relationships

- Deafblind people don't fit anywhere
- Friends withdraw
- Community roles/activities are difficult - attendance stops
- Transportation is difficult
- Asking for help can engender embarrassment
- Deafblind person is marginalized by their identified group
- The deafblind person can become more isolated, withdrawn and grief-stricken.

Vocational

- Accommodation only goes so far
- People leave/lose their jobs early
- Further loss of self esteem, identity, roles
- Job retraining– perhaps with less remuneration
- Fewer people work as their vision deteriorates

Employment 87 adults, Usher I, 1998, US

- 41 people, ages 21 – 30: 53% were working or in school
- 19 people, ages 31 – 40: 47% were working or in school
- 9 people, ages, 41 – 50: 33% were working
- 18 people, ages 52 – 60: 22% were working or at university-
these should be prime working years-
The three who were working have postgraduate degrees, were literate in English and use Braille. The university student is also literate in English and uses Braille.

Adaptation/Acceptance

- Don't like word "acceptance"
- Usher isn't the same disease every day or year
- Progression requires ongoing repeated adaptation and change
- Issues change.. New solutions are always necessary

Grief and mourning

- Grief is not linear
- Grief is not a one time process
- Grief is cyclical & recurrent
- Grief reaction for person with Usher and family

Resurgence of Grief- Person with Usher and family

- There is further vision or hearing loss.
- Normal developmental events don't happen on time
- There are unrelated losses or major life events
- New accommodations have to be made.

Denial

- Much maligned- but it allows hope
- Purpose: protect integrity of the individual
- Unconscious mechanism
- Functional vs. dysfunctional denial
- If it's functional, support it- 'I will learn these skills even though I'll never lose my vision.'

It isn't easy to ask for help

- Applying for service is an acknowledgement of loss.
- Can feel shame, guilt, like a burden
- Further blow to self esteem, sense of identity, feelings of independence and competence.
- The result can be depression, grief, withdrawal, and disequilibrium in the family

These losses sound very grim, but

Outcomes differ depending on available choices

Denmark's system of "contact persons" - alleviates isolation

In Seattle, WA a similar system of support service providers.

In New York City, there is no such system

These services allow communities to flourish and people to develop a sense of deafblind identity.

Rehabilitation/New skills

- Orientation and Mobility
- Braille to maintain literacy
- New household skills
- Vocational training or retraining and work
- Equipment and adaptation
- Contact persons or SSPs- SSPs not available everywhere
- All contribute to a sense of competence and mastery over what is happening

The skills no one wants to talk about



These are signs of independence, not dependence

Reconstituting identity

- Positive role models, folks further down the Usher road
- Contact with peers with Usher
- The availability of a real community
- Contact persons or support service providers to be able to take part in that community and maintain independence.
- Productive activity & enjoyment in the community

Some attributes of deafblind culture

From a Danish woman:

We care about each other, one person talks at a time, we are aware of each other's needs, we check on communication, we don't pass judgment, there is a lot of touch, and a strong sense of belongingness, and among each other, we don't have to explain ourselves.

The pace is slower, life takes planning, sign and spoken communication changes

Summer camp



Danes and New Yorkers

- Everyone spoke of the importance of meeting and being with others with Usher, that this was often the beginning of progress.
- Differences between Denmark and New York: sense of Identity and Community. “I am deafblind”
- Deafblind Danes have more options and opportunity to come together to form a community
- Seattle WA has options and SSPs- very different from NY

Being with others who are deafblind

- “I feel normal.”
- “I feel like I have come home.”
- “I see that deafblind people can lead normal lives.”
- I don't have to explain myself; we understand each other
- When asked to identify their identity, most of the Danes responded, “I am deafblind”. The New Yorkers did not.

Some fun– photo from Norway



Having a chat



The Danes

- Most identified as deafblind
- The Danes had more options to be together.
- Identification as deafblind not dependent on degree of hearing or vision loss.
- Community and culture are inextricably woven with sense of identity
- All those who self-identified as deafblind participated in national deafblind association (FDDB) and in regular ERFA groups

The New Yorkers

- Only two of the group called themselves ‘deafblind’
- There is no sense of community or place where deafblind people go to be together
- There is no established support service provider program that would make this possible
- The difference between New York and Seattle is stark

The US

- No right in the law to the services of support service providers
- Only 17 programs around the US
- Fewer hours per deafblind person than in Denmark
- Deafblind community in Seattle
- It is the only established community, smaller communities do exist with fewer services.

St Patrick's Day Party, CA



The world is full of equipment



More equipment

