



## Human and Social Sciences research on daily life and life courses with Usher syndrome

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*LIGHT4DEAF*

**RECHERCHE HOSPITALO-UNIVERSITAIRE EN SANTÉ**

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### PARTNERS



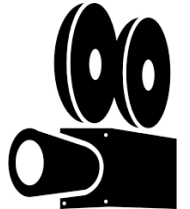
# Integrative Method



- Qualitative interviews (213):  
71 concerned ADULTS (29 US1, 35 US2, 1 US3, 6 ?)  
40 healthcare professionals, specialists and non-specialists  
50 everyday professionals (social, linguistic, educational, cultural)  
52 relatives: parents, children, spouses



- Two sociological intervention groups: Usher patients and entourage  
10n people from the Paris region and the provinces  
6-7 work sessions of 3-4 hours; several accessibility methods



- Data crossing:
  - “UsherSHS” questionnaire on life courses with deafness and vision difficulties 266 responses including 172 US (52 US1, 91US2, 6 US3, 22 ?)
  - 1 demo-sociological info sheet: characteristics of the survey population
  - Observations: consultations, daily life , associative life ...

## Diagnostics and Announcements

- Several diagnostics and Announcements:

Announcement of deafness (vestibular tbl) made to parents during childhood, diagnosis of evolution of hearing loss

Announcement of clinical diagnosis either presymptomatically or when visual clinical signs appear

Announcement of visual field narrowing

Announcement of genetic diagnosis

- The moment of the announcement can be a relief or a real traumatic shock

almost 2/3 had no psychological follow-up at the time of the announcement

1/2 declares that they did not understand the information at the time of the announcement

2/3 did not receive adequate help and 1/3 had suicidal thoughts

1/4 are currently in psychological distress

- Awkward announcements

Awareness issue for non-specialist ophthalmologists ("you are going to go blind")

Avoid associating types of syndrome with severity levels of visual impairment

Words to talk about genes: "bad", "defective", "to fix", "with errors"

- Difficulties in telling and accompanying the evolution

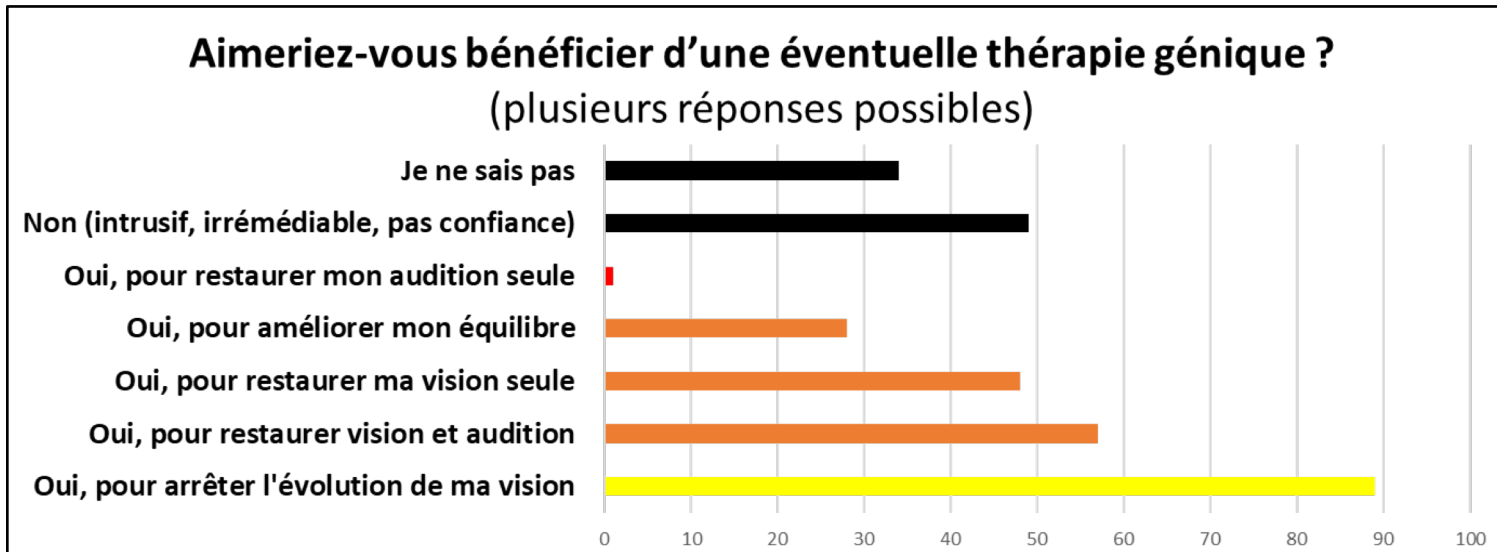
Talking about a cane is received as an announcement of loss of sight

The cane, linked to total blindness in words, and not in practice

# Follow-up and support following the diagnosis announcement

- ❑ Giving meaning to the experience, integrating the syndrome into the identity, providing keys to know how to express your needs
- ❑ Follow-up offered with genetic diagnosis ... at a distance from clinical diagnosis
- ❑ The follow-up does not only concern the patient: need to take care of relatives (first helpers: exhaustion, guilt, stress)
- ❑ Learn to deal with negative emotions: 50% use negative coping strategies (eg 25% straight or self-aggressive aggression, 36% isolation); Appropriateness of acceptance and commitment therapy follow-up
- ❑ The follow-up is useful over time, with other interlocutors providing solutions in ordinary life, interest in training psychologists in the practices of deafblind people

## Expectations with respect to medical therapy



Need information on possible treatments (74%)

45% received information about gene therapy from their doctor

Don't know if they received the information (13%)

- Their medical problem concerns vision
- Deafness, as a norm, because it is already integrated into their identity and their daily lives
- Blocking the evolution of vision: the feasibility? the reliability of the therapies?
- The refusal to consider treatment is not heresy, because it is hypothetical and in tension with the work of psychological acceptance
- Access to alternative strategies is as difficult as it is vital

# Information Needs

- Informations about health

For 83%, doctors are the source of health information

Some people have difficulty to understand the difference between types of syndrome

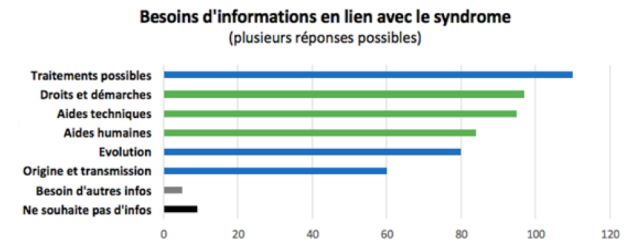
Need for information on the origin and heredity (34%) and the course of the syndrome (48%)

Need for rapid debriefing of medical check-ups and hospital conferences

- Informations on human and technical aids

more important than the need for health information

- technical aids (62%)
- possible human assistance (58%)
- rights and procedures (67%): complex administrative procedures
- need for a resource person to centralize administrative procedures
- need to be put in contact with specialized associations
- lack of awareness of the diversity of professionals and their contributions



# Complexity: understanding everyday experiences

- Importance of adjustments according to daily contexts
    - Variability of needs according to places, environments, etc. : eg. Cane
  - Contextual and situational blindness: "I am a blind person who sees" (according to activities, tasks, time, etc.)
  - Expectations concern
    - Management of communication conditions (distance, mobility, noise, etc.)
    - Organization of domestic space, storage and associated fatigue
    - Help in the entourage too
  - Requests from relatives:
    - Access to the diversity of information: care, techniques, daily life
    - Understanding the references, experiences and needs of the Usher person
    - Knowing what to do, how to do it: without suffering, imposing, or replacing
    - Find their place
- ➡ Issue of organizing this support

# Take into account the variability of practices

- 1/2 of “deafblind” people and 1/2 cane users:  
Say they see or see “badly”  
Have an objective visual field greater than 10 °
  - ❖ Contextual uses of the cane: darkness, blinding light, crowd, attention paid to something other than its movement (children, etc.)
- The “deafblind” are 1/3 of tactile sign language users; 3/5 of the people use bilingual, bimodal (Oral / lip reading / written / Sign Language /Tactile)
  - ❖ People at risk: Sign Language with poor level of written French and poor tactile communication skills; Lip reading + no auditory rest + no SL + poor tactile skills

# Identity work in face of the evolution of visual loss

- Challenge: find a stable and positive way to respond to whatever may happen (adjustment, capacity and interaction)

## ❖ Different strategies:

Renuncements so as not to be seen otherwise, withdrawal and disaffiliation

Integration into networks sharing alternative practices (deaf signing, blind, deafblind)

Identity anticipation, positive because it is controlled and decided by themselves

Valuation and self-construction around an ability to reposition oneself (driving temporarily, changing jobs, places of life, etc.)

- Challenge: be able to freely access information and assistance

Whenever you want

On his initiative: to know the networks and the contacts

And unfettered

## Socially constructed obstacles

- Discrimination by ignorance and negative anticipation ("he will not be able")

By some health professionals, training, working, but also parents and spouses (worried and anxious)

Do not give access to the different modes of communication : Fear and depreciation of codes involving touch; Lack of knowledge of the possibilities of SL and its tactile version

Do not facilitate immediate access to training and professional experience due to future visual impossibilities

Over-invest in protection (refuse independent journeys, certain sports, responsibilities)

Relatives who do not project themselves into a possible life with deafblindness

- ❖ Avoid a "corridor logic" and open up the field of possibilities: Consider positive anticipation, Encourage a diversity of experiences, Encourage sport, Do not anticipate limitations

## Work and Access to Rights

- Work: very present and central, although nervously tiring

Difficulties in finding a job: others do not project themselves

Diversity of professions but high rate of "inactive"

1/3 do not have the necessary aids

1/3 consider work without impact or good for health, but > 1/2 have work-related nervous fatigue

- Administrative and political non-recognition of deafblindness

General approach based on mono-deficiency and on medical criteria

Deafblindness defined by two handicaps of high severity (profound deafness + legal blindness)

Ignorance of the practical difficulties of moderate-severity combinations

Not taking into account an anticipation of strategies (transition phases)

❖ Consequences: exhaustion and identity shattering

# Take Home Messages

- INFORM: ophthalmologists, psychologists, general practitioners of the specifics of the care of deafblind people
- CENTRALIZE information: creation of a register bringing together associations specializing in deafblindness, psychologists, social workers, interpreters and the need for medical reports
- SUPPORT the process of adaptation to the disease: at each diagnosis to allow the disease to be integrated into the patient's history, use of active cognitive techniques offering concrete solutions
- PROMOTE alternative solutions and positive anticipations: appropriation of the cane, of tactile sign language upstream blindness



Thank you for your attention !

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<http://ushersocio.org/resultats.html>