Landau-Kleffner syndrome in Norway

Long term prognosis and experiences with the health services and educational system

Nordic conference on rare diseases – Reykjavik Iceland 2012
Epilepsy related disorders
Landau-Kleffner syndrom

- Described by Landau and Kleffner in 1957 (Smith, 2003)
- Affecting children in the age between 3-8 years (Nickels 2008, Panayiotopoulos 2008)
- Increase of epileptic activity during sleep (Nickels 2008, Panayiotopoulos 2002)
- Epileptic activity longer than 2 - 3 years, increase neuropsychological sequelae (Robinson 2001, Smith/Hoepner 2003, Soprano, 1994)
- Approximately 70-80 % of the children have epileptic seizures (Paquier 1992, Duran 2009, Msall 1986)
- Controlled trials of treatment is lacking (Smith, 2003)
- Behavioral disturbances is often an important component of the syndrome (Smith, 2003)
- We know 30 people with Landau- Kleffner in Norway (Bølling, 2005)
Landau-Kleffner syndrome in Norway

- Purpose
- Material and methods
Demographic data for patients diagnosed with Landau Kleffner syndrome (n=19)

<table>
<thead>
<tr>
<th></th>
<th>Sex</th>
<th>Age at diagnosis</th>
<th>Age at onset of aphasia</th>
<th>Age at first symptom</th>
<th>Time from first symptom to diagnosis</th>
<th>Time from onset of aphasia to diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>6 female</td>
<td>13 men</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Age at diagnosis</td>
<td></td>
<td>Mean 6,2 years</td>
<td>Min: 2,8 years</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Median 5,5 years</td>
<td>Max: 12,4 years</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Age at onset of aphasia</td>
<td></td>
<td>Mean 3,7 years</td>
<td>Min: 1,4 years</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Median 3,4 years</td>
<td>Max: 6 years</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Age at first symptom</td>
<td></td>
<td>Mean 2,9 years</td>
<td>Min 0,5 years</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Median 2,8 years</td>
<td>Max:10,9 years</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Time from first symptom</td>
<td></td>
<td>Mean 3,3 years</td>
<td>Min: 0,8 years</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Median 2,2 years</td>
<td>Max: 10,9 years</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Time from onset of aphasia to diagnosis</td>
<td>Mean 2,5 years</td>
<td>Min: 0,3 years</td>
<td>Max: 11,0 years</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
The ICD 10-criteria of LKS

- Normal language development followed by loss of impressive and expressive language
- Preserved general intelligence
- Paroxysmal abnormalities in EEG at debut of symptoms
- Age of onset between 3 and 7 years
- Epileptic seizures present in most cases
- Exclusion criteria are unspecific language disturbances, disintegrative disorders, and autism
Language disruption

- 7 auditory agnosia
- 2 verbal agnosia
- 10 auditory discrimination deficit
<table>
<thead>
<tr>
<th>Epilepsy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Epileptic seizure</td>
</tr>
<tr>
<td>Seizure types</td>
</tr>
<tr>
<td>CPS</td>
</tr>
<tr>
<td>6 (32)</td>
</tr>
<tr>
<td>Age at first seizure</td>
</tr>
<tr>
<td>Median/min-max</td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td>Age seizure free</td>
</tr>
<tr>
<td>Median/min-max</td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td>Years of seizure</td>
</tr>
<tr>
<td>Mean/median/min-max</td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td>Seizure free today</td>
</tr>
<tr>
<td>Number (percent)</td>
</tr>
</tbody>
</table>
Behavioral, attention and social abilities

- 17 children were described as exhibiting behavioral patterns of frustration or aggression
- 7 children had physically behavioral patterns
- 16 children had difficulties in their interactions with others and in exhibiting appropriate behavior
- 14 children were described as having concentration or attention-deficit problems
- 5 were suspected of having a hyperactivity disorder
- 11 had mood fluctuation
- Short-term memory was studied at follow-up for all patients considered suitable for testing (12 cases) and was found to be impaired in all 12
Long term outcome

Language

- 4 normal verbal language
- 4 moderate language problems
- 3 no functional verbal language
Long term outcome

Work/study

Five of our patients have now reached adulthood

- four in paid employment
- one is studying

Three of them have completely verbal language, and two have moderate language difficulties
Parents first concern

- Language decline: 58%
- Late language development: 26%
- Seizure: 11%
- Loss of contact: 5%

Average age at symptom onset: 2.9 (range 5 months-11 yrs)
Professionals initially contacted

- General practitioner: 37%
- Nursery school staff: 16%
- Psychologist/educator: 21%
- Local hospital: 16%
- Public health nurse: 11%
Professionals mentioning the diagnosis at the first time

- Pediatrician: 53%
- Family member: 12%
- Psychologist student/internship candidate: 29%
- Doctor at the Norway National Epilepsy center: 12%
Where was the diagnosis made?

- 42% referred to a local/regional hospital and got the LKS diagnose
- 32% referred to a national epilepsy center because of possible LKS and got the diagnose
- 26% referred to a national epilepsy center because of epilepsy and got the LKS diagnose
Diagnostic delay

For those with

- auditory discrimination deficit: 4.1 years (SD 3.6)
- auditory/verbal agnosia: 1.5 years (SD 0.9)
To be listened to

”They said it was nothing wrong with him. You lose confidence when they do not listen to what you say”
(father)

Delays in diagnosis and inappropriate interventions cause rare disease patients to lose confidence in the healthcare system and in medical professionals
(Kole, 2009)
Consequences of the diagnosis

- More appropriate help
- Better understanding
- Parents could do something to help
- Hope
- Being listened to
Contributions from the professionals
Parents would like

- Less struggle
- Leading coordinator
- Better adjustments
- More knowledge
- Taken seriously
- Reliability
Summery

- There was considerable heterogeneity in the children’s symptoms
- Late onset language decline, short duration of the initial language problems and marked fluctuations in speech abilities associated with a positive outcome
- Diagnose contributed to more appropriate help and better understanding of the child
- Many were not taken seriously when they expressed their worries, and they expressed a strong wish for someone who could ensure that appropriate support was implemented
Thanks!

- The children and their parents who participated in the study
- Supervisor Karl Otto Nakken
  National center for Epilepsy, Oslo University Hospital
- Supervisor Hilde Robinson
  Institute of Health and Society, University of Oslo
- Grete Bølling
  Solberg School, County Administration, National Center for Epilepsy, Oslo University Hospital
- The Norwegian Directorate of Health for financial support
Thank you for your attention!
References

Smith MC, Polkey CE. Landau-Kleffner syndrome and CSWS. In: Engel J jr., Pedley TA, editors, Epilepsy. A comprehensive textbook. 2nd ed. Philadelphia: Lippincott Williams & Wilkins;2008:2429-37
World Health Organization. ICD 10: Mental disorders and behavioral disturbances: clinical characteristics and diagnostic criteria. Oslo: Universitetsforlaget; 1999