PROGNOSTIC FACTORS IN PAEDIATRIC COCHLEAR IMPLANTATION (PCI)

Description Distribution Evaluation

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BACKGROUND 1

Why prognosticate in PCI?

- The concept of prognosis in PCI has not yet attained clinical acceptability and credibility.
- Prognostication is difficult – importantly the devil is in the detail.
BACKGROUND 2
Achieving an accurate prognosis

- Clear recognition of any adverse factors that may impede an optimal PCI outcome
- Have the ability to develop an accurate cumulative evaluation of these factors
BACKGROUND 3

There are three aspects relevant to the prognostic process in PCI

1. **Description**: a clear definition of prognostic threats to the PCI process: *What are they?*
2. **Distribution**: Where will they act along the auditory pathway?
3. **Evaluation**: *What is the impact* of the individual and cumulative prognostic factors / threats present within each case
1. THREAT DESCRIPTION

Past Difficulties

- Use of Aetiology, e.g. “Meningitis”, “CMV” etc.
- Ill-defined terms, e.g. Inner Ear Malformations
- Ambiguous terms, e.g. Age at Implantation
- Inaccurate description, e.g. “Auditory Neuropathy Spectrum Disorder” vs. true VIII nerve pathology

To date, little focused literature
Example 1
“Meningitis”
Outcomes of Well vs. CNS-damaged Cases

Meningitis cases: Well children vs. Children with damaged Central Nervous System (CNS)

<table>
<thead>
<tr>
<th>Case Outcomes</th>
<th>Well children vs. Children with damaged CNS</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Excellent  Good  Fair  Poor  Not Available  Total</td>
</tr>
<tr>
<td>Well Child</td>
<td>6     1   -  6*    2   15</td>
</tr>
<tr>
<td>CNS</td>
<td>-    -    3    -     -    3</td>
</tr>
<tr>
<td>Total</td>
<td>6    1    3    6     2   18</td>
</tr>
</tbody>
</table>

Note. *3 Cases of Family Difficulties and 3 Cases of Ossified Cochlea

i.e. the pathology was relevant, not the aetiology
Example 2
“Inner Ear Malformations”

A. LVA: Good prognosis

B. Cochlear dysplasia: Suspect prognosis

Better focus needed
THREAT DESCRIPTION

Summary

- Previous poor scrutiny of the relevant threats
- Inadequate focus on pathology degraded the quality of the data used
- More precise evaluation of prognostic threats is needed
2. THREAT DISTRIBUTION

1. Identify the exact mechanism of the site and nature of the pathological effect on the auditory pathway.

2. Understand the action of the threat/s on pathway function from CI to cortex.

Visualise where the threat is and how will it impact on ability.
THE AUDITORY CORTEX
Role in Prognostication

- The cortex is the ultimate site of success or failure in PCI
- Cortical function (and thus the prognosis) may be degraded in two ways
  - Damage: to the cortex itself
  - Denial of Data
    - Impeded maturation
    - Interruption of input
Anatomical Distribution of Threats to Cortical Ability (PCI Success)

Primary effects:
- Auditory cortex
- Neurological

Secondary effects:
- Cochlear
- VIII Nerve

Tertiary effects:
- General Medical
- Psychological
- Family / Social

Illustration courtesy Robert Jackler MD

n = 60 cases of 174

20% of cases were multifactorial
EFFECT OF CORTICAL THREATS

Six Domains Affected

- **Primary Influences**: Cortical damage
  - Cortical maturation – delayed PCI
  - Neurological damage

- **Secondary influences**: Failed transmission
  - Otological: Cochlea, VIII

- **Tertiary Influences**: Failed PCI stimulus
  - General Medical
  - Psychological
  - Family / Social (environmental)
3. THREAT EVALUATION:
Individual and Cumulative Effect
Prognostic Assessment Process

- Study isolated prognostic factors
- Multiple factors when present may span several domains
- How do we do this? A prognostication method has been developed
# Prognostication Method

1. **Identify individual threats** and list each factor in the relevant domain affected

2. **Assess Individual Threat Probability**
   - The clinically assessed **likelihood** that the threat will affect the PCI outcome:
     - 1. Minimal
     - 2. Possible
     - 3. Probable
     - 4. Certain

3. **Assess Individual Threat Severity**
   - Clinical estimation of the **severity** of the perceived threat:
     - 1. Minimal
     - 2. Slight
     - 3. Substantial
     - 4. Severe

4. **Overall Individual Threat Assessment**
   - Likelihood $\times$ Severity. The scores are then re-classified, 1-4, minimal to maximal risk:
     - 1. Minimal threat: 1-4
     - 2. Some threat: 6, 8
     - 3. Considerable threat: 9, 12
     - 4. Major threat: 16

5. **Individual Domain Rating**
   - Threats within the domain are tabled. **Combined effect** is calculated 1-4, minimal to maximal, using the three assessments listed below.
     - Rate according to worst threat
     - Two “3” threats = 4
     - Three “2” threats = 3

6. **Combined Domain Rating: Prognosis**
   - Domains are tabled, **assess prognosis** as per prior box, 1-4
     - 1: Excellent prognosis
     - 2: Good prognosis
     - 3: Fair prognosis
     - 4: Poor prognosis
## Case Prognostication:
### Combined Domains Assessment (Prognosis)

<table>
<thead>
<tr>
<th>Domain</th>
<th>Factor</th>
<th>Threat Probability</th>
<th>Threat Severity</th>
<th>Overall Threat Rating</th>
<th>Comment</th>
<th>Domain Rating</th>
</tr>
</thead>
<tbody>
<tr>
<td>Auditory Cortex</td>
<td>CMV damage</td>
<td>3</td>
<td>2</td>
<td>2</td>
<td>Some threat</td>
<td>2</td>
</tr>
<tr>
<td>Neurology</td>
<td>MRI – CMV changes</td>
<td>3</td>
<td>3</td>
<td>3</td>
<td>Considerable threat</td>
<td>3</td>
</tr>
<tr>
<td></td>
<td>Poor motor skills</td>
<td>3</td>
<td>2</td>
<td>2</td>
<td>Some threat</td>
<td></td>
</tr>
<tr>
<td>Otology</td>
<td>Nil</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>Minimal threat (EABR normal)</td>
<td>1</td>
</tr>
<tr>
<td>General Medical</td>
<td>Nil</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>Minimal threat</td>
<td>1</td>
</tr>
<tr>
<td>Psychology</td>
<td>Reduced intellect</td>
<td>3</td>
<td>2</td>
<td>2</td>
<td>Some threat</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Delayed milestones, poor cognition</td>
<td>3</td>
<td>3</td>
<td>3</td>
<td>Considerable threat</td>
<td>3</td>
</tr>
<tr>
<td>Family / Social</td>
<td>Disruptive family</td>
<td>3</td>
<td>3</td>
<td>3</td>
<td>Major threat</td>
<td>4</td>
</tr>
<tr>
<td></td>
<td>Poor compliance</td>
<td>4</td>
<td>3</td>
<td>3</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
SPECIALISED TOOLS
Prognostication and Auditory Performance Measurement

1. Prognosis: Family Assessment Profile
2. Prognosis: Cochlear Implant Paediatric Prognostic Index (CIPPI)
3. Outcomes: Categories of Auditory Performance Index (CAPI)
FAMILY ASSESSMENT PROFILE

- Derived from the Nottingham Children's Implant Profile (NChIP)
- Used to classify the possible impact of the family situation on both prognosis and outcome.

Units of analysis

- Family structure / support
- History of hearing loss / impact on family / child
- Understanding of CI process / family expectations
- Compliance / commitment aspects

Graded: 1 = No Concern; 2 = Mild Concern; 3 = Great Concern
CIPPI
A pathological and social compilation of perceived threats assessed in the 6 domains

- It provides a framework for the evaluation of isolated or multiple adverse factors based on the literature (evidence-based)
- Evaluation is specific to the child
- Provides an orderly pathway of assessment
CAPI

- An index of performance categories arranged in increasing ability based on the child’s performance on tests of auditory perception.
- Nine descriptive categories ranging from nil or limited awareness to advanced open set speech perception in noise on auditory perception test performance.
- It succeeds the Nottingham Categories of Auditory Performance (CAP).
- It is objective, repeatable and a valuable research tool.
CONCLUSION

- PCI prognostication requires precise evaluation of the site, pathology and action of adverse threats / factors evident in a case.
- Focus on the specific pathology of the threats.
- Avoid vague terminology / descriptors.
- When assessing a PCI prognosis, examine the auditory pathway systematically for evident prognostic threats.

Each case is a clinical judgement; cases will vary with individual situations.