Single Event Multilevel Surgery (SEMLS) for Children with CP: The evidence from Europe and Australia

Pam Thomason, Jessica Pascoe, Kerr Graham
Hugh Williamson Gait Analysis Laboratory & Department of Orthopaedics, Royal Children’s Hospital, Melbourne.

Martin Gough
One Small Step Gait Laboratory, Guy’s Hospital, Evelina London Children’s Hospital, St Thomas’ Hospital, London.

Contributors: Tim Theologis, Oxford, UK and Eric Rutz, Basel, Switzerland.

Definition and classification of cerebral palsy

Cerebral palsy describes a group of permanent disorders of the development of movement and posture, causing activity limitation, that are attributed to non progressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, perception, cognition, communication, and behaviour, by epilepsy, and by secondary musculoskeletal problems.

Musculoskeletal problems

- Contracture of muscle-tendon unit
- Bony torsion deformity
- Joint instability – hip, mid foot

Musculoskeletal problems

- Progressive
- Decline in motor function and gait

Surgical management: single level, multiple occasions

Birthday Syndrome: Mercer Rang

<table>
<thead>
<tr>
<th>TAL</th>
<th>H/S</th>
<th>Poses</th>
<th>Rectus femoris</th>
</tr>
</thead>
<tbody>
<tr>
<td>Squat</td>
<td>Crouch</td>
<td>Flexion</td>
<td>Stiff knee</td>
</tr>
</tbody>
</table>
What is SEMLS?
- Single Event Multilevel Orthopaedic Surgery
- Major intervention
- Multiple procedures in one surgical intervention
- Correction of all fixed deformities
- Requires comprehensive assessment
- Requires comprehensive evaluation of outcome
- Primarily for improvement in Gait with potential to improve function and quality of life
- Requires a team approach to management
- Intensive rehabilitation

SEMLS
- Part of an overall management plan for the individual child from 2 to 18 years

Orthopaedic surgical management and goals of surgery is dependent on:
- Severity and function (GMFCS)
- Goals of child and family
- Multidisciplinary team involvement

SEMLS evaluation
- Diagnostic Matrix

Single Event Multilevel Surgery: SEMLS
- Soft Tissue
- Bony
SEMLS procedure

**Soft tissue**
- Psoas
- Hip adductors
- Hamstrings
- Gastrocnemius
- Rectus femoris

**Bony**
- Femur
- Tibia
- Foot

Correct levers: Transverse Plane

Pre SEMLS  3 years Post SEMLS

Surgical dose: Knee

Surgical Dose: Gastrocsoleus

Measuring Outcomes

- Movement Analysis Profile
- Gait Profile Score
Movement Analysis Profile (MAP) and Gait Profile Score (GPS)

Gait Variable Score (GVS) average (RMS) difference between gait trace and average curve for children with no pathology for each kinematic variable

Gait Profile Score average of 9 GVS for left, right and overall

MAP and GPS comparison

Movement Analysis Profile
BBF 2009-01-27

Movement Analysis Profile
BBF 2010-08-10

Movement Analysis Profile
BBF 2014-02-10

Evidence for SEMLS

- Systematic Review

SEMLS Evidence

- Systematic review McGinley 2011
- 31 studies
  - Level of evidence
    - 23 Level 4, 7 Level 3 (control), 1 Level 2 (post op rehab)
  - Majority (18) retrospective
  - Outcomes mainly body structure & function
    - Gait
      - 8 studies improvement in temporospatial parameters
    - 16 studies improvement in kinematics
  - Activity
    - 2 studies improvement in GMFM, 4 unchanged

Evidence for SEMLS
Evidence for SEMLS
- RCT with 5 year follow up

RCT and 5 year follow up
- Aim RCT
- To evaluate the effect of SEMLS on:
  - Gait
  - Motor function
  - Mobility
  - HR QoL
- Follow up study

Study Design
- RCT
- Follow up study
- Baseline & randomisation
- SEMLS
- Control/PRST
- Proceed to SEMLS
- Both groups combined
- 1 yr post SEMLS
- 2 yr post SEMLS
- 5 yr post SEMLS

Participants
- Inclusion:
  - 19 participants RCT
  - Spastic diplegia
  - GMFCS Levels II (n=14) & III (n=5)
  - Mean age 9.4y
- 5 year follow up
  - 18 participants
  - Age range 2.8 to 18.2y
  - One child excluded
    - Diagnosis revised from CP to HSP

Surgical procedures
<table>
<thead>
<tr>
<th>Level, type</th>
<th>No.</th>
<th>Mean/Child</th>
<th>No. subsequent</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hip</td>
<td>73</td>
<td>4</td>
<td>1</td>
</tr>
<tr>
<td>Knee</td>
<td>25</td>
<td>1</td>
<td>9</td>
</tr>
<tr>
<td>Ankle / Foot</td>
<td>44</td>
<td>2</td>
<td>12</td>
</tr>
<tr>
<td>Soft Tissue</td>
<td>97</td>
<td>5</td>
<td>9</td>
</tr>
<tr>
<td>Bony</td>
<td>45</td>
<td>3</td>
<td>5</td>
</tr>
<tr>
<td>Growth Plate</td>
<td>0</td>
<td>0</td>
<td>8</td>
</tr>
<tr>
<td>Total</td>
<td>143</td>
<td>7</td>
<td>22</td>
</tr>
</tbody>
</table>

Results follow up
- 0 to 5 years
  - Mean height increase 29.7 cm
  - Mean weight increase 25.1 kg
Results - GPS

![Bar chart showing improvement maintained with p=0.0001](chart1.png)

Results - GMFM

![Bar chart showing 3.3% improvement maintained](chart2.png)

Results - CHQ

![Bar chart showing improvement maintained with p=0.01](chart3.png)

Functional Mobility - FMS

- 5yrs - pre surgery

![Frequency change 5yrs - pre surgery chart](chart4.png)

Results - CHQ

Conclusions SEMLS in CP

- SEMLS safe and effective correction musculoskeletal deformities
- With functional rehabilitation
  - large sustained improvements in gait
  - small sustained improvements in gross motor function
- Improvements in HRQoL
- Improvements across all ICF domains

Longer term evidence for SEMLS

- Cohort Studies
  - Hinden et al, ESMAC 2013
Cohort Studies

- Retrospective: Level IV evidence
- Prospectively gathered standard data
- Small and moderate subject numbers
- Real world data
- More generalisable than RCT?

Svehlik et al, 2011

- Aim
  - The effect on age at time of surgery on long term outcome following SEMLS

Results

- 32 children
- Mean age 10.6 (3.1)y
- GMFCS II & III
- Follow up 1 to 10 years post SEMLS
- Outcome GDI
- Surgery required at older age do better than at younger age


Zwick et al, 2012

- Aim
  - Does gender influence the long term outcome following SEMLS

Results

- 34 children
  - 19 boys and 15 girls
- Mean age boys 9.9, girls 11.3y
- GMFCS II & III
- Follow up 10 years post SEMLS
- Outcome GGI
- Both groups improved post surgery but the boys deteriorated in the long term


Gait Outcome studies

- Two studies
  - Hinden et al ESMAC, 2013
- Number of children
  - Rutz 121
  - Hinden 160
- 2 year follow up
- Outcome GPS
Single event multilevel surgery in Switzerland

Erich Rutz 1,2,3
Kerr Graham 2

1 Pediatric orthopaedics, University Children’s Hospital Basel UKBB, Switzerland
2 Murdoch Childrens Research Institute, Royal Children’s Hospital, Victoria, Australia
3 Laboratory for Gait Analysis, University Children’s Hospital Basel UKBB, Switzerland

Introduction

Switzerland...

SWISS SEMLS study

• Published in Clin Orthop Relat Res, 2013; Rutz et al.
• Small cohort (n=14) with severe gait dysfunction
• Are results after SEMLS durable?

Material & Methods

14 patients with spastic diplegia
- 4 girls, 10 boys; mean age: 12.8 ± 3.3 years
• GMFCS
  - 1x level I, 10x level II, 3x level III
• SEMLS
  - 6.4 surgical interventions per session, range: 4-10
  - bony and soft-tissue procedures
• Preoperative, short- and mid-term 3DGA

Results: pelvis and hip kinematics

Results: knee and ankle kinematics

After 2 years

After 5 years

After 2 years
Results: MAP & GPS

Conclusions:

• Short term: Severe gait dysfunction can be improved in one operative session (SEMLS)

• But, to preserve the early improvements two third of the patients required additional surgery (relapses or new gait problems)

Outcomes Predictors for Multilevel Surgery in Children with Cerebral Palsy/Spastic Diplegia

Introduction

• Overall, despite good results in Multilevel surgery, the selection of candidates can still be challenging.

• There is a need for better criteria to predict individual outcomes

• Rutz, et al. ESMAC 2012
  - SEMLS outcome after 1 year
  - Pre-op GPS
  - Age at surgery
  - Surgical adverse events

Methods

• Retrospective Cohort Study, 1994 – 2012

• 160 spastic diplegic cerebral palsy patients (age 6 to 24 yrs; GMFCS I-III) who received SEMLS

• Gait analysis pre-op and approximately 2 years post-op
  - Conventional lower body model

• The GPS* for each gait analysis was calculated separately with an Excel® template


Results

• Overall:
  - 75% improved (MCID 1.6°) *Baker
  - 2 % deteriorated (MCID 1.6°)
  - Average improvement of 3.2°
Results

- Outcome Predictors:
  - Pre-op GPS was found to be the strongest predictor of change ($b=0.57$, $p<0.001$) and was also significantly associated with absolute GPS post-op ($p<0.001$).
  - Meaning: higher GPSs are related to better result.

- Other Outcomes:
  - Age at surgery was found to be a significant predictor of change ($p=0.02$), but not absolute GPS following surgery ($p=0.61$). Younger patients
  - Bilateral vs Unilateral bilateral showed greater change ($p=0.001$)
  - Soft tissue only vs bony +/- soft tissue not a significant predictor of change ($p=0.60$) or outcome ($p=0.37$).

Conclusion

- Similar results to Melbourne point towards the applicability of results throughout different centres.
- Pre-op GPS strong predictor of SEMLS outcome
- Age was a predictor determined by multiple regression
- Including surgery classification in the analysis did not improve the prediction. Further studies needed.
- The change in the GPS (preop – postop) was sustained up to 2 years following surgery

- GPS Improvement = (0.6 x pre-op GPS) – (0.1 x age) – 2.6
### Results: Gait Profile Score (GPS)

- Improvement
  - GPS 4.3° (Rutz)
  - GPS 3.2° (Hinden)
- MCID = 1.6° *
- Most severe gait improved the most
- Pre op GPS strongest predictor of improvement
- This may be helpful in decision making?


### What have we learnt?

- Age and gender may effect outcome
- Deformity correction is immediate
- Gait correction at 1 year
- Functional changes take longer (2 years)
- Medium term follow up
  - Not everyone improves why?
  - Is improvement maintained long term?
- Gait analysis can measure outcome
- Gait analysis can predict outcome?

### Background

- Hereditary Spastic Paraparesis (HSP)
  - gait disturbance
  - lower limb spasticity
  - weakness
- HSP similar to but important differences from CP
- Research
  - ability to identify HSP as different from CP using 3DGA
- Comparison with CP difficult
  - wide variety of presentation
  - severity of impairments

### Hereditary Spastic Paraparesis (HSP)

- How this differs from CP
- Principles of SEMLS for HSP

### HSP – functional differences

### Results – Sagittal gait patterns in HSP

<table>
<thead>
<tr>
<th>Gait pattern</th>
<th>n</th>
<th>Pelvis</th>
<th>Hip</th>
<th>Knee</th>
<th>Ankle</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sagittal Mild</td>
<td>9</td>
<td>Variable</td>
<td>Normal flexed</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>True Equinus</td>
<td>9</td>
<td>Variable</td>
<td>Normal flexed</td>
<td>Normal</td>
<td>Recurvatum</td>
</tr>
<tr>
<td>Jump</td>
<td>4</td>
<td>Variable</td>
<td>Normal flexed</td>
<td>Fixed</td>
<td>Fixed</td>
</tr>
<tr>
<td>Crouch</td>
<td>2</td>
<td>Variable</td>
<td>Fixed</td>
<td>Fixed</td>
<td>Calcaneus</td>
</tr>
<tr>
<td>Asymmetric</td>
<td>2</td>
<td>Combination of any two of above patterns</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Results:
- HSP 80% full knee extension
- BSCP 30% full knee extension

Summary
- Gait in HSP shows variability as in BSCP
- More children with HSP demonstrated full knee extension in gait compared to BSCP
- New ‘sagittal mild’ classification lowest median GPS indicating closer to normal gait

Clinical Implications
- For this cohort increased knee extension has implications for surgical dosing
  - BSCP – Over lengthening of plantarflexors = crouch
  - HSP - Protection against recurvatum with hamstring lengthening?
- Distribution of impairments may differ to BSCP
  - Dominance of calf spasticity?
  - Compensation will also be different
  - Balance between spasticity and weakness

To treat or not to treat?
- Main impairments spasticity and weakness
- No joint contractures
- Significant gait abnormalities
Conclusions

- There are differences in sagittal gait patterns between HSP and BSCP
  - New sagittal mild no apparent equinus
  - In particular – knee extension in stance
  - This may contribute to protection against crouch in the long term and hence progressive disability

Gait corrective Surgery in HSP

- Use the clinical algorithm as in CP
- Research shows gait is improved and maintained in CP
- What about HSP?

Retrospective Cohort study

- 10 children
- 3DGA pre surgery and post surgery at T1 – 12 months 12-24 months and T3 4-6 years

<table>
<thead>
<tr>
<th>GMFCS level</th>
<th>n</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>2</td>
</tr>
<tr>
<td>II</td>
<td>5</td>
</tr>
<tr>
<td>III</td>
<td>3</td>
</tr>
</tbody>
</table>

Results:

- 7/10 showed clinically significant improvements in GPS at T2 and T3 (MCID = 1.6)
- No statistical significance – Heterogeneous cohort

Results: Medium GPS at short term and medium term 3DGA

- Majority of children gait was maintained or improved
- Severe gait impairments = greater improvements
- Mild gait impairments = less evidence of change
- Appropriate to use 3DGA, surgical intervention and intensive rehabilitation as per protocols in CP for HSP
SEMLS RCH

- Surgery Protocols: emphasis on “dose”
- PIP funded rehab and 3 monthly monitoring is the RCH point of difference
- Inpatient rehab (VPRS)
- How does that work, in practice?

SEMLS RCH

- SEMLS rehab lasts 12-24 months
- Early mobility
- Need for monitoring, evaluation, feedback, & corrective steps
- Small team, with instant communication
- Gait Lab Physios and Surgeons
- SEMLS/PIP Physio
  - Post op assessment & reporting
  - CP nurse coordinator
  - Communication
- Community Physio/Family/Child
- VPRS
  - Multi disciplinary approach
  - In patient rehab
  - Long term follow up

SEMLS and the Gait Lab

- Pre op 3DGA surgical decision making
- Monitor rehab progress, 3, 6 9 months
- Outcome evaluation 3DGA at 12 months
- Removal of hardware, “fine tuning”
- Outcome evaluation 3DGA at 24 months
- Monitor progress 2D or 3D as appropriate
- Outcome evaluation 3DGA at 5 years
- Transition to adult care & long term follow up