Brachial Plexus

Traumatic injury:
- Terminal branches
- Ventral rami

From Netter 1997
Upper Brachial Plexus Injuries
- Increase in angle between neck & shoulder
- Traction (stretching or avulsion) of upper ventral rami (e.g., C5, C6)
- Produces Erb’s Palsy

Lower Brachial Plexus Injuries
- Excessive upward pull of limb
- Traction (stretching or avulsion) of lower ventral rami (e.g., C8, T1)
- Produces Klumpke’s Palsy

“Obstetrical” or “Birth palsy”
- Becoming increasingly rare
- Categorized on basis of damage
  - Type I: Upper (C5,6), Erb’s
  - Type II: All (C5-T1), both palsies
  - Type III: Lower (C8, T1), Klumpke’s Palsy

From Moore & Dalley (1999)
Upper Brachial Plexus Injury: Erb’s Palsy

- Appearance: drooping, wasted shoulder; pronated and extended limb hangs limply (“waiter’s tip palsy”)
- Loss of innervation to abductors, flexors, & medial rotators of shoulder and flexors & supinators of elbow
- Loss of sensation to lateral aspect of upper extremity
Lower Brachial Plexus Injury: Klumpke’s Palsy

- Much rarer than UBPIs and Erb’s Palsy
- Loss of C8 & T1 results in major motor deficits in the muscles working the hand: “claw hand”
- Loss of sensation to medial aspect of upper extremity

From Moore & Dalley (1999)

From Netter 1997
Case Presentation

A 5-year-old boy is brought to the pediatrician with the complaint that since early childhood the right side of his neck has been twisted and deformed. Childbirth apparently had been prolonged and difficult and was a breech delivery. Within a few weeks, there was a spindle-shaped swelling on the right side of the neck that was tender on touch and on passive movement of the head. Over the next few months, the swelling and tenderness subsided. By the time the boy was about 1-year-old, the muscle on the right side of the neck appeared cordlike. Gradually the neck became stiff and deformed, as shown at left. The face was also asymmetrical.

From Moore & Dalley (1999)

Case modified from Cahill (1997)
Congenital Muscular Torticollis

- Fibromatosis colli that develops in SCM probably prior to birth, although birth trauma (e.g., forceps) has also been implicated.
- 75% of cases on right side.
- SCM is transformed into a cordlike, nonfunctional muscle, distorting head and neck posture and altering growth of the face.
- Etiology is unclear: Arterial or venous obstruction? Intrauterine malposition?
Sternocleidomastoid

- Attachments: proximally, mastoid proc. & occ. bone; distally, sternum & clavicle
- Innervation: accessory n., C2,3

- Surgical concerns
  - nerves emerging from post. border (esp. accessory n.)
  - jugular veins
  - carotid A. & its branches

- Bilateral contraction: flex the neck
  - pull chin toward sternum
  - in conjunction with neck extension: protrusion of the chin

- Unilateral contraction: ipsilateral neck flexion and contralateral rotation of chin (as in torticollis)

From Moore & Dalley (1999)
Asymmetries Secondary to Congenital Muscular Torticollis

- Ipsilateral shortening and flattening of face; ipsilateral depression of eye & ear
- Contralateral convex scoliosis in lower cervical and upper thoracic regions; compensatory ipsilateral convex scoliosis in middle & lower thoracic regions

Photo from Tachdjian (1990)
“Pulled (Nursemaid’s) Elbow”

- Very common in children under 4 (peak: ages 1–3)
- More in common in boys; more common on left side
- Sudden traction with elbow extended & forearm pronated

From Netter 1987
Subluxation of the Radial Head (“pulled elbow”)

- Annular lig. joins radial head to ulna
- Traction on pronated forearm causes a distal tear in the annular ligament where it merges with the periosteum
- Radial head “escapes” anteriorly
- Annular ligament slides onto articular surface of radial head, between radial head & capitulum

- Above the age of five, tear does not occur because of thicker attachment of annular ligament to periosteum
Why Subluxation in Pronation?

- Annular ligament restrains radial head, while allowing axial rotation of the radius
- Steep & sharp lip anteriorly between radial head & neck—in supination, annular ligament cannot slip proximally over lip
- Shallower lip laterally and posteriorly
- During pronation, rotation of radius brings this shallow lip anteriorly, allowing proximal slip of ann. lig. if there is a transverse tear
Interposition of the Annular Ligament

- Extent of interposition of the annular ligament within the joint determines course
- If ligament does *not* extend beyond “equator” of radial head (B below), subluxation can be reduced by closed manipulation (passive supination)
- If ligament extends beyond equator (A below), open (surgical) reduction may be required
“Breaking News”

Self-Actuating Extendible Endoprosthesis

- Prosthesis designed for children requiring knee replacement (due to malignant bone tumors)
- Previous implants required repeated surgery to adjust for growth and to avoid leg-length discrepancy
- New device does not require repeated surgery: it extends automatically with flexion of the knee

From Kotz et al. 2000
References


Biological regeneration of the extremities can occur in some organisms (starfish, for example), but humans need recourse to a mechanical solution. Growing children present a particular challenge, and those who lose bone tissue from the knee after removal of a tumour need an implant that can not only provide stability for the femur and allow motion of the knee joint, but which also accommodates rapid growth. Here we describe a self-extending leg implant (endoprosthesis) that can closely simulate natural growth and which has worked successfully in paediatric patients. The energy needed for elongation of this device is provided by the patients themselves as a result of flexure of the knee joint, thereby reducing the number of operations and the risk of infection associated with manually extended implants.

The outlook for patients with malignant bone tumours has historically been poor, but now increased sophistication in medical imaging and adjuvant treatment has dramatically improved survival. The success of limb-sparing surgery has also stimulated technological developments in designing and manufacturing endoprostheses to restore function to the damaged limb.

Although bone implants have proved effective in adults, they cause a significant limb-length discrepancy which precludes their use in growing children. Extendible prostheses have been developed to match the growth of the opposite limb, but frequent surgery was necessary for elongation procedures, for aseptic loosening of the growing bone, and to correct failure of the extension mechanism. In a long-term follow-up of un cemented endoprostheses implanted after tumour resection of the lower leg in 13 out of 44 children with growth arrest, the final clinical and radiographic results matched those of similarly treated adult patients. Invasive methods involving manual, stepwise elongation of the implant also cause scar formation that limits motion of the knee joint.

Despite their drawbacks, the application of these earlier extendible implants proved successful in 13 patients, with definitive limb salvage and equal limb length after growth was complete. These are compared in Table 1 with two patients treated with the endoprosthesis, in which continuous automatic elongation is brought about by enforced knee bending.

Overlengthening of the self-extensible prosthesis is prevented by the tension of the surrounding soft tissues, which increases after each elongation procedure (0.056 mm) and is gradually decreased as the soft tissue grows, thereby allowing the knee more flexure and stimulating the elongation mechanism. The number of surgical procedures needed for the flexion-controlled device has been reduced (Table 1). A further improvement for children under treatment now is the introduction of a central camshaft into the implant (Fig. 1), which allows the knee to bend painlessly beyond the point of elongation (100-degree bend) up to an angle of 140 degrees.

**Table 1** Comparative performance of manually and self-actuating extendible endoprostheses

<table>
<thead>
<tr>
<th></th>
<th>Manual device</th>
<th>Self-actuating device</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at resection of tumour (yr)</td>
<td>11.4 (7.3–16.1)</td>
<td>9.3 (6.5–12.1)</td>
</tr>
<tr>
<td>Age at implantation of module (yr)</td>
<td>12.8 (10–18)</td>
<td>12.9 (10.3–15.5)</td>
</tr>
<tr>
<td>Deficit in leg-length at implantation (mm)</td>
<td>72 (5–40)</td>
<td>–48 (–45–51)</td>
</tr>
<tr>
<td>Total lengthening after implantation (mm)</td>
<td>84.8 (11–196)</td>
<td>157 (145–169)</td>
</tr>
<tr>
<td>Step lengthening (mm)</td>
<td>50–100 mm, by surgery</td>
<td>0.138 mm d⁻¹</td>
</tr>
<tr>
<td>Number of operations*</td>
<td>10 (5–25)</td>
<td>7.5 (6–9)</td>
</tr>
<tr>
<td>Follow-up (months)</td>
<td>112.6 (70.6–158.1)</td>
<td>105.2 (100.7–109.7)</td>
</tr>
</tbody>
</table>

*Includes surgical procedures for implantation complications.

**Figure 1** Function of the self-actuating extendible endoprosthesis (known as an intercondylar stepless-extension module). Flexure of the knee joint induces clockwise rotation of a switch unit (a). An angle of flexion of 100 degrees moves a gear wheel (b) via a carrier by 20 degrees; this positioning is held by a spring (c). Rotation of the gear wheel is translated to a threaded spindle (d), which in turn causes a movement of 0.056 mm in a telescopic sleeve higher up the leg (e). An 18-fold repetition of this movement causes an elongation of 1 mm.
Insect metabolism

Preventing cyanide release from leaves

Organisms that produce hydrogen cyanide gas to protect themselves against predators can do so by the enzymatic breakdown of a class of compounds known as cyanogens (such as cyanogenic glycosides). Here we show how a neotropical butterfly, Heliconius sara, can avoid the harmful effects of the cyanogenic leaves of Passiflora auriculata (passion vine), on which its larvae feed exclusively. To our knowledge this is the first example of an insect that is able to metabolize cyanogen and thereby prevent the release of cyanide. The mechanistic details of this pathway might suggest new ways to make cyanogenic crops more useful as a food source.

Many organisms, including humans, possess enzymes for detoxifying hydrogen cyanide (HCN). Barriers against herbivory of cyanogenic plants may be more effective as a result of the slow or inefficient detoxification by the herbivore of HCN or of byproducts of the enzymatic breakdown of cyanogenic glycoside, which generate not only HCN but also toxic aglycones upon ingestion. Nonetheless, cyanogenesis is no defence against specialist herbivores such as the larvae of Heliconius butterflies. We investigated how Heliconius butterfly larvae process cyanogenic glycosides found in their Passiflora host, a diverse neotropical genus in which coevolution with Heliconius accounts for morphological innovation in some species.

We examined the fate of five classes of Passiflora cyanogens ingested by Heliconius larvae by surveying the distribution of cyanogens found in 12 species of Passifloraceae and 14 species of teneral adult Heliconius reared on specific hosts. We extracted cyanogens from fresh Passifloraceae and butterflies and identified them using published procedures.

All butterflies contained aliphatic cyanogens, irrespective of their presence in host plants, extending previous findings that these cyanogens are synthesized de novo by Heliconius. Most cyanogen classes in hosts were not retained in the butterflies, nor extracted in larval frass. However, we found monoglycoside cyclopentenyl cyanogens in all butterflies fed on Passiflora containing these compounds. Acraea horta, a distant relative of Heliconius, is known to sequester a cyclopentenyl cyanogen from Kiggelaria plants, but we have now demonstrated cyanogen sequestration in Heliconius.

Figure 1

**Figure 1** Structures of sequesetered cyanogen from *H. sara*. **a**, Epivolkenin, and **b**, the corresponding thiol derivative sarauriculatin (epivolkenin thiol). Spectral data for epivolkenin: (15.49–1-bp-o-glucopyranosyl)-4-hydroxy-2-cyclopentene-1-carbonitrile. *'H NMR (250 MHz, CD3OD) signals at 8 6.14 (dd, H-2), 6.27 (dd, H-3), 4.81 (m, H-4), 2.25 (d, H-5A), 3.02 (dd, H-5B), 5.6 Hz, 3J5'-6'A = 5.5 Hz, 2J5'-6'B = 12.0 Hz, 1J5'-6'B = 5.5 Hz, 1J5-A = 1.3 Hz, 1J5-B = 2.0 Hz, 1J6-A = 14.8 Hz, 1J6-B = 4.8 Hz, 1J7-A = 7.7 Hz, 1J7-B = 4.6 Hz, 2J2,4 = 7.3 Hz, 1J5-A = 1.3 Hz, 1J5-B = 4.6 Hz. 1J4,5A = 13.9 Hz, 1J4,5B = 6.4 Hz, 1J5',6'A = 13.9 Hz, 1J5',6'B = 2.0 Hz, 1J6',7' = 7.3 Hz, 1J5,6 = 4.6 Hz, 1J6,7 = 7.3 Hz, 1J7,8 = 7.5 Hz. 1J1,2 = 3.2 Hz, 3J1,2 = 8.3 Hz, 3J1,6 = 8.3 Hz, 3J2,3 = 8.3 Hz, 4J2,3 = 13.9 Hz, 1J1,6 = 13.9 Hz, 1J1,7 = 13.1 Hz, 4J1,2 = 6.2 Hz, 3J7,8 = 8.3 Hz, 4J7,8 = 13.1 Hz, 1J1,6 = 13.9 Hz, 1J1,7 = 13.1 Hz, 4J1,2 = 6.2 Hz, 3J7,8 = 8.3 Hz, 4J7,8 = 13.1 Hz, 1J1,6 = 13.9 Hz, 1J1,7 = 13.1 Hz, 4J1,2 = 6.2 Hz, 3J7,8 = 8.3 Hz, 4J7,8 = 13.1 Hz, 1J1,6 = 13.9 Hz, 1J1,7 = 13.1 Hz, 4J1,2 = 6.2 Hz, 3J7,8 = 8.3 Hz, 4J7,8 = 13.1 Hz, 1J1,6 = 13.9 Hz, 1J1,7 = 13.1 Hz, 4J1,2 = 6.2 Hz, 3J7,8 = 8.3 Hz, 4J7,8 = 13.1 Hz, 1J1,6 = 13.9 Hz, 1J1,7 = 13.1 Hz, 4J1,2 = 6.2 Hz, 3J7,8 = 8.3 Hz, 4J7,8 = 13.1 Hz, 1J1,6 = 13.9 Hz, 1J1,7 = 13.1 Hz, 4J1,2 = 6.2 Hz, 3J7,8 = 8.3 Hz, 4J7,8 = 13.1 Hz, 1J1,6 = 13.9 Hz, 1J1,7 = 13.1 Hz, 4J1,2 = 6.2 Hz, 3J7,8 = 8.3 Hz, 4J7,8 = 13.1 Hz. 'H NMR of sarauriculatin: 

Paper chromatography and high-pressure liquid chromatography of cyanogens from *H. sara* and its host plant *P. auriculata* were followed by specific enzyme tests, [H- and 13C-NMR analyses, and mass spectrometry. We compared spectral data from these cyanogens and their tetracyclililane-ether derivatives with published spectra for cyanogens of Passifloraceae and related families. The primary cyanogens in *P. auriculata* were monoglycoside cyclopentenyl cyanogen, epivolkenin (90%), its diastereomer taraktophyllin (5%) and a diglycoside cyclopentenyl cyanogen (5%). Of these, only epivolkenin was sequestered by *H. sara* (Fig. 1a).

We isolated a previously unknown metabolic derivative of epivolkenin, sarauriculatin, from *H. sara*, which we identified as ([1S,4R]-1-(4-Hydroxy-2-cyclopentene-1-thiol). Spectral data for epivolkenin: (1S, 4R)-1-(4-hydroxy-2-cyclopentene-1-carbonitrile (Fig. 1b). Sarauriculatin was present at a ratio of 1:2 with epivolkenin. This new compound was characterized as a cyclopentenyl cyanogenic glycoside that has undergone replacement of the nitrile group by a thiol, a metabolic reaction that pre-empts the enzymatic release of cyanide.

Conversion of nitrile groups is due to specific enzymatic action, and conversion of cyclopentenyl cyanogenic glycoside nitriles to their amides occurs spontaneously in plant isolates, probably as a result of mixing with enzymes. In repeated tests, however, we found sarauriculatin in butterfly but not in plant isolates. We conclude that a unique enzymatic mechanism exists in *H. sara* for dealing with cyanogenic glycosides.

Our results show that cyanogen sequestration in *H. sara* must be both selective and dynamic, because only one type of host cyanogen is sequestered and this is metabolized to release valuable nitrogen into the insect’s primary metabolism. A fuller understanding of this process should reveal its importance in the nutrition of some species of Heliconius, and provide a new perspective on *Heliconius–Passiflora* coevolution, and suggest how agriculturally important cyanogenic plants might be rendered safer and more nutritious for human and animal consumption.

Helene S. Engler*, Kevin C. Spencer†, Lawrence E. Gilbert*

*Section of Integrative Biology, School of Biological Sciences, University of Texas at Austin, Austin, Texas 78712, USA
e-mail: h.engler@mail.utexas.edu

†Maytomer Manor, Wendover Dean, Buckinghamshire HP22 6QA, UK

5. Søger, D. S. & Brüker, A. M. in Methods in Plant Biochemistry

© 2000 Macmillan Magazines Ltd