Welcome to Texas
Principles And Practice For PHOTOPHERESIS

Presented by Regina Mack, RN., HP(ASCP)
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Apheresis Department
Off-label” and investigative uses of drugs and devices will be discussed.

The speaker has no relevant financial obligations with vendors.
Outline

• Introduction
• UVADEX
• History
• Machine Technology
• Clinical Indications:
  • Cutaneous T-Cell Lymphoma (FDA approved)
  • Graft Versus Host Disease acute and chronic (off-label)
  • Solid organ rejection (off-label)
  • Crohn’s disease (off-label)
  • Multiple sclerosis (off-label)
• Types of access
• FACT-JACIE International Standards for Photopheresis
• Insurance
Extracorporeal photopheresis (ECP) is a type of immunomodulatory therapy that harvests a leukocyte enriched buffy coat via centrifugation. The cells are treated outside the body with an injectable solution called UV ADEX, a psoralan-based medication, exposed to Ultraviolet lights, then reinfused to the patient. Upon photactivation, there’s a conjugation and formation of covalent bonds with DNA strands which leads to the formation of monofunctional and bifunctional adducts.¹ This process is believed to interact with proteins and subsequently results in apoptosis, which suppresses T-cell mediated conditions but does not produce an inflammatory response. The treatment is used for cellular rejection as well as other disease processes.

There are many theories to the understanding. However, a full understanding of the process is somewhat a mystery.
Apoptosis is a form of programmed cell death that does not result in an inflammatory process. Whereas necrosis cell death is due to injury that results in inflammation.
Mechanism of Action

Possible mechanism of ECP

Regina’s theory on mechanism of action

Phagocytosis

Macrophage

Apopotic Cell

Retrieved from Kangle@Deviantart.com
UVADEX

Comes from the seeds of the Ammi Majus plant with a history that dates back as early as 2000 BC in Egyptian times.

Ammi Majus—also known as Bishop’s weed, Queen Anne’s, bullworth, lace flower, meadow sweet, etc.
History

Richard Edelson, MD ’70, Aaron B. and Marguerite Lerner Professor of Dermatology and chair of the department at Yale, received the 2015 Discovery Award during the annual meeting of the Dermatology Foundation on March 21 in San Francisco.

- Identified and named the disease known as cutaneous T-cell lymphoma (CTCL)
- Developed the first FDA-approved therapy
- Elucidated principles of tumor progression and resolution

Allen Latham Jr. also known as “Jack” was an industrial engineer and founder of the Haemonetics Corporation in 1971. He is credited with the invention of the “Latham Bowel”.

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Richard Edelson, MD ’70 receives discovery award. Retrieved from Yale news http://medicine.yale.edu/dermatology/

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First FDA Approved Machine
FDA APPROVED MACHINES

UVAR XTS

TherakosCellex

http://www.omniamed.net/en/prodotto/cellsaver-5-plus.html

Courtesy of Therakos

Courtesy of Therakos inc
Instrumentation

**UVAR XTS**
- Single Needle discontinuous
- Duration 2 ½ – 4 hours
- Heparin
- Closed System
- Latham bowel
- 2 bowel sizes (125ml and 225ml)
- ECV 220-620mls
- Minimum HCT 28
- No minimum WBC
- Requires manual calculation for UVADEX dose
- Ability to return blood manually
- Manual ability to pack red cells with pause during buffy coat

**Therakos Cellex**
- Double or single needle and switch between the two
- Duration 1 ½ to 4 hours
- Choice of Heparin or Citrate
- Closed System
- Latham bowel (modified)
- Shorter prime times
- One bowel size
- Smaller ECV (220 double-216 single)
- Can be used on patients 22kg(requires blood prime
- Minimum HCT 27
- No minimum WBC
- Ability to return blood manually
- On screen assistance with alarms
- Smart card
- Smaller footprint
- Manually pack red cells with pauses during buffy coat
ASFA Categories as of 2013

I  Disorders which apheresis is accepted as a first line of therapy as primary stand alone or in conjunction with another type of therapy.

II  Disorders for which apheresis is accepted as a second line therapy as a stand alone treatment or in conjunction with another mode of therapy. ECP with corticosteroids

III  Optimum role of therapy is not established. Decision making should be individualized. ECP

IV  Disorders which published evidence demonstrates or suggests apheresis to be ineffective or harmful. Institutional review board approval is desired if treatment is undertaken.

Grading Recommendations from Guyatt et al.

Grade 1A Strong recommendation. Can apply to patients in most circumstances without reservation

Grade 1B Strong recommendation. Can apply to patients in most circumstances without reservation

Grade 1C Strong recommendation but may change when higher quality evidence becomes available

Grade 2A Weak recommendation best action may differ depending on circumstances

Grade 2B Weak recommendation best action may differ depending on circumstances

Grade 2C Very weak recommendation; other alternatives may be equally reasonable

Transfusion 2014
Clinical Indications

- Cutaneous T-Cell Lymphoma (CTCL) FDA approved diagnosis
  - Sezary Syndrome
  - Mycosis Fungoides
- Graft versus Host Disease (GVHD)
  - Acute and Chronic
- Allograft lung rejection
  - Bronchiolitis obliterans syndrome (BOS)
- Allograft cardiac rejection
- Other solid organ rejection
  - Face
  - Liver
  - Kidney
- Multiple Sclerosis
- Crohn’s Disease
- Atopic dermatitis
- Type 1 Diabetes
- Pemphigus
- Epidermolysis bullosa acquisita
- Erosive oral lichen planus
- Lupus erythematosus
- And many more
Procedural concerns

Hypotension
Dizziness, lightheadedness
Access problems
Pain or bruising at needle insertion site
Do not exceed more than 15% of patient total blood volume
Hematocrit
Weight
Decision between double or single needle
Keep patient warm during procedure
Anemia
Elevated bilirubin
Chills
Paresthesia
Contraindication

- Pregnancy or lactating women
- Reaction to psoralan medications
- Must be used cautiously in patients receiving PUVA treatments
- Use of systemic or topical medications that are known to be photosynthesizing
- Procedure is not indicated for patients that have a disease that causes them to be photo sensitive to certain artificial lights or UV rays eg. Xeroderma, or Erythropoietic Protoporphyria (EPP), Aphakia disorders
- Weight less than 45kg
- HCT less than 27
- Heparin use with patients with history of Heparin Induced Thrombocytopenia (HIT)
Vascular Access

Procedure can be managed via central line or peripheral accesses
- Peripheral Access 16 or 17G AVF needle
- May Return via 18G Angiocath
- Quinton Catheter (temporary)
- Hickman
- Permacath
- Neostar
- Tri-fusion (UVAR XTS)
- Vortex
- Arteriovenous fistula or graft
- Power PICC?
- Sport Port

Without access there is no treatment and with poor access there is often times inadequate treatment

Pre-Procedural Communication/Practice

- Hydrate few days prior to procedure
- Consume low fat meals
- Light fluid intake on the day of procedure
- Start wearing 100% UVA protected sunglasses on day 1
- Know what medications patient is taking.

- Collect a CBC prior to procedure
- Collect a CBC from treatment bag duringuffy coat.
- Measurement of collection efficiency by fold enrichment
- Patient’s medical history
- Weight checks
1. Estimated ECV weight in KG X body build factor ml/kg = TBV
   Maximum Safe ECV range TBV multiply by 0.10 and 0.15 = ECV range

2. Units of Heparin delivered 10,000/500mls X AC volume = heparin given

3. UVADEX dose: TV X 0.017 = UVADEX ml

4. Measurement of collection efficiency
   Fold enrichment:
   Product or hemogram WBC/ Baseline WBC = fold enrichment

**Goal:** is to have at least a “1” as measured by fold enrichment
Example: product wbc 10/ pre wbc 5 = 2
Discharge Communication/Concerns

• Photosensitivity
• Possible urticaria (redness and itching)
• Hypertension
• Temperature elevation
• Fatigue
• Anemia – for patients with more treatments than the recommended amount
• Patients should be advised to follow treatment plan as ordered
• Response times vary between patients
• Use sunscreen with a minimum of 15% SPF
• Post procedure bleeding or complications
• Line care
ALARMS!!!!


Call and report to Therakos technical support

http://bauscharddebate.com/2013/02/11/overcoming-conflict-winning-through-teamwork/
Request for more specific guidelines (C8.17)
A written therapy plan from a physician
✓ Diagnosis and GVHD grade, involved organs, indication, timing of procedure, proposed regimen etc.
✓ Final report of ECP
✓ Documented agreement between transplant physician and Apheresis physician.
✓ Must have educational material and initial consultation occurred prior to therapy.

Standard operating procedure
Final report of ECP administered

FACT-JACIE 6th Edition Standards

Insurance Concerns

Not all insurance will cover the cost of Photopheresis.

• Pre-authorization or pre certification may be required depending on insurance company.

• Letter of medical necessity
Access Program

Administered for Therakos through a joint venture with the National Organization for Rare Disorders (NORD).

• Available by request only and to those that qualify

Contact information:
www.rarediseases.org
therakos@rarediseases.org

1-877-241-7220
Monday – Friday
9am to 5pm EST
DONT MESS WITH TEXAS!