

# Exstrophy

**The Bladder Is *Abnormal*  
at Birth**

# Why CPRE Early ?

**Evidence of  
Unique Potential  
for Bladder  
Healing with  
early CPRE, but  
may be time  
limited**

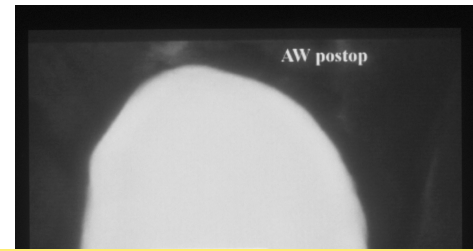


# Why the EARLY AND COMPLETE?

- **Clinical Observation;**
  - **Posterior Urethral valves**



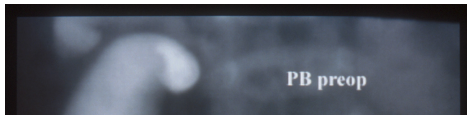
# Retrospective Study



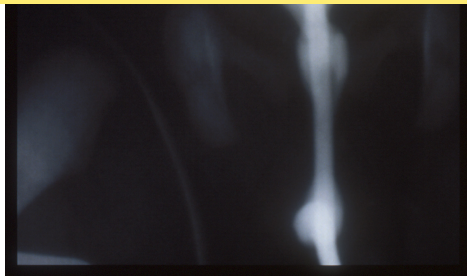
Better results if urine **not Diverted** from Bladder



**Early** Valve  
Ablation, only



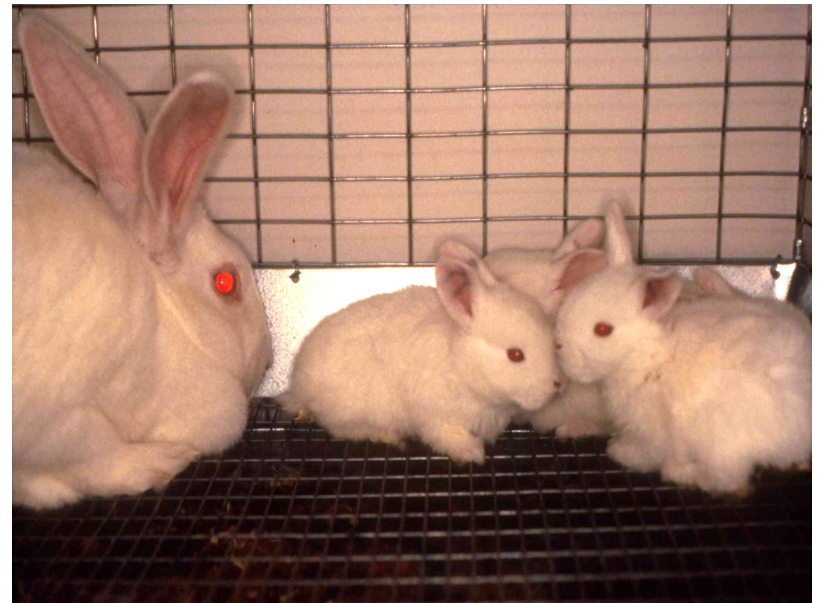
The bladder can heal with Normal Filling and Emptying, but this must be very soon after birth



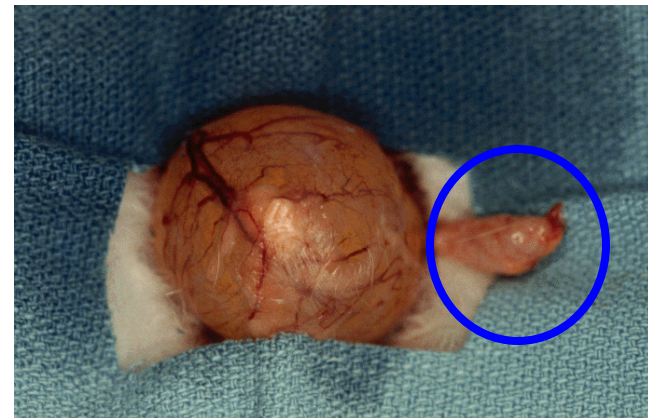
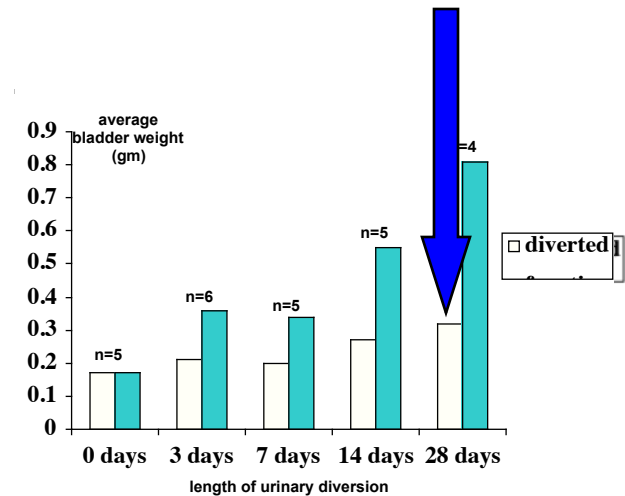
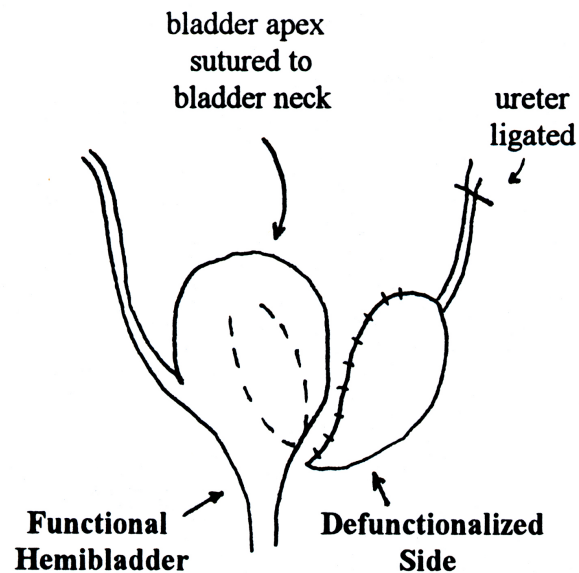


# Experimental Studies

- **Show the Unique Physiology of the Neonatal Bladder**
- **Rabbit Model**
  - **Obstruction**
  - **Diversion**



# Experimental Diversion

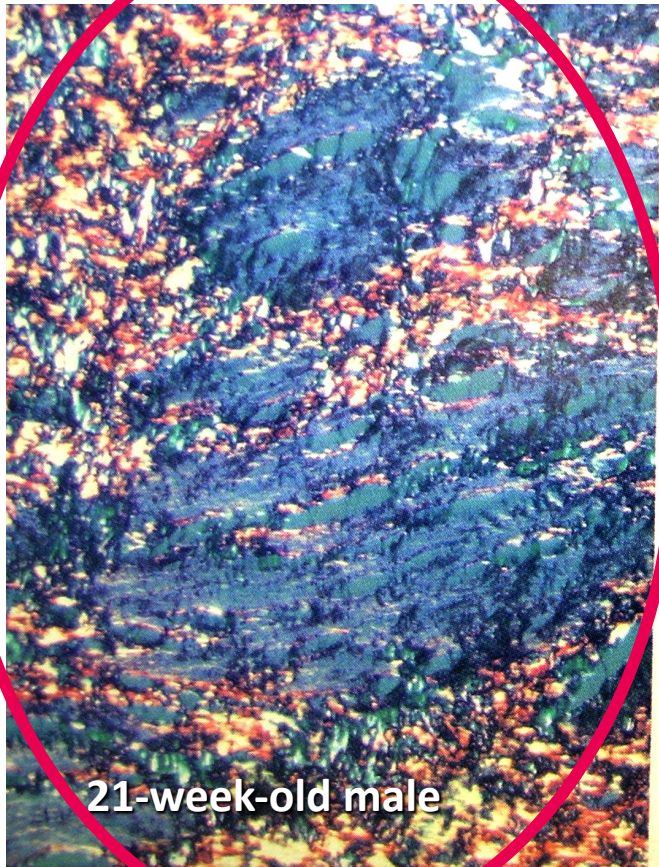




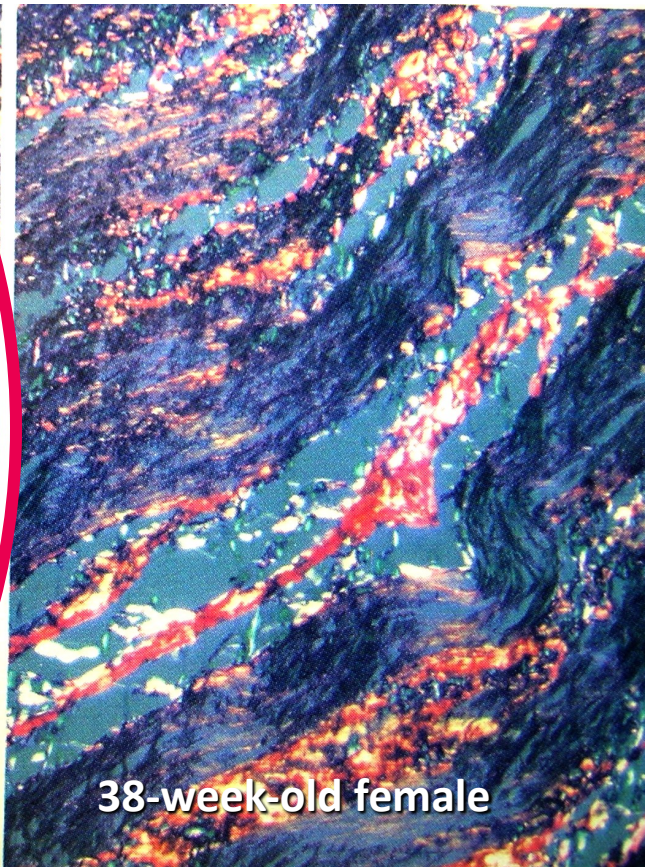


# Normal Fetal Bladder

Polarizing light microscopy and Sirius red stain of fetal bladder\*



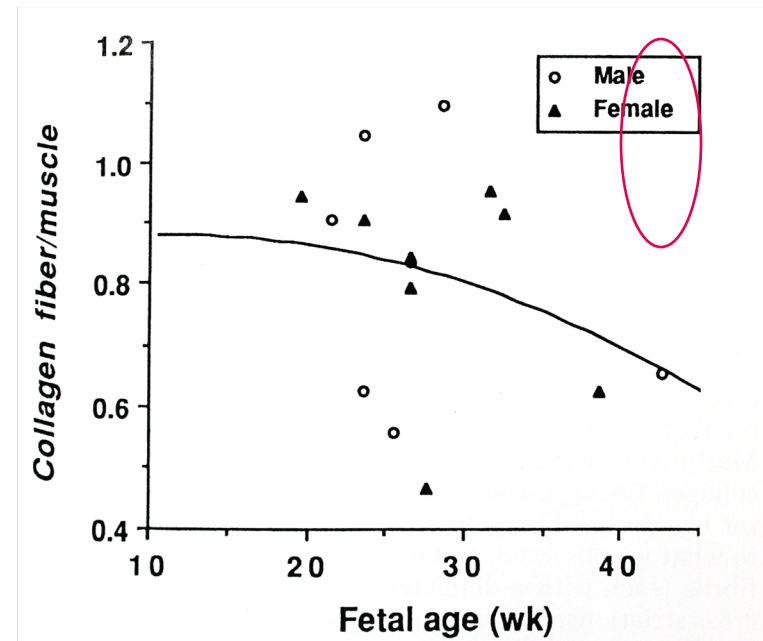
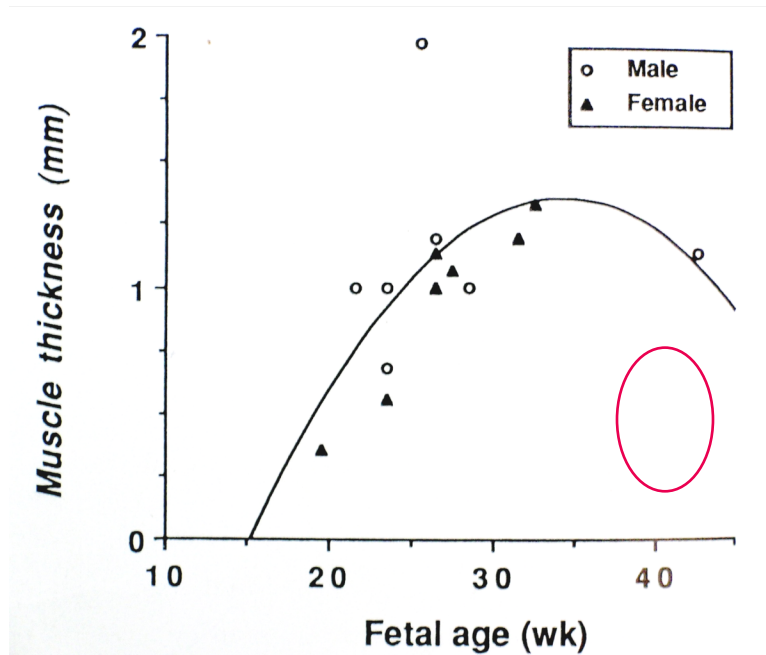
21-week-old male



38-week-old female

\* Kim, KM, et al. J Urol 146, 525, 1991

# Exstrophic Bladder at birth may be “stuck” at an early phase of Development\*



**Exstrophic Bladder Collagen/smooth muscle is 2.5x of control\*\***

\*Kim KM, et al., J Urol, 146, 524-527, 1991

\*\*Lee BR, et al., J Urol, 156, 2034-2036, 1996

# Why One Stage at Birth? (Developmental)

## **Window of Opportunity**

The Bladder has Unique Properties  
for Development and Healing in the  
First Few Months of Life.

Clinical observations (valves, exstrophy,  
etc.)

Research (consistent with Developmental  
Biology)



# Exstrophy Is a Surgical Problem

As surgeons, all we can do is  
rebuild the anatomy

And hope **function** will follow form

# Etiology

**“Single Hit”** → **Hole** → **Herniation**

## TIMING OF EVENT

**Early**

**Claoca (4-7wks)**



**Cloacal**

**Later**

**After tailbud  
after Kidneys**



**Classic**

**Latest**



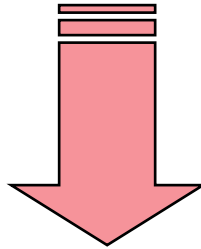
**Epispadias**

# Exstrophy

# Why One Stage at Birth?

## Hypothesis:

**“Normal” Filling and Emptying (Cycling)  
of the Bladder**



**Normal Bladder Development**

# Continence in Exstrophy Children

- Therefore there is Good Potential for bladder volume and Compliance if---
- Primary Closure results in Bladder Cycling (i.e., CPRE)

# Key points for today

1. **Exstrophy is a spectrum**
2. **Exstrophy bladder is abnormal at birth**
3. **Abnormal Bladder can 'normalize' if cycled**

# Long-term results

## Bladder Neck and Urethra, Genitalia

1. Voiding with Continence
2. Sexual Identity/Function
3. Fertility

When??

Spectrum dependent



