



VENOLYMPHATIC MALFORMATION in a 4 year old
male treated with **PICIBANIL (OK-432)**
with **SYSTEMIC INFLAMMATORY RESPONSE SYNDROME**
as a complication

A CASE REPORT

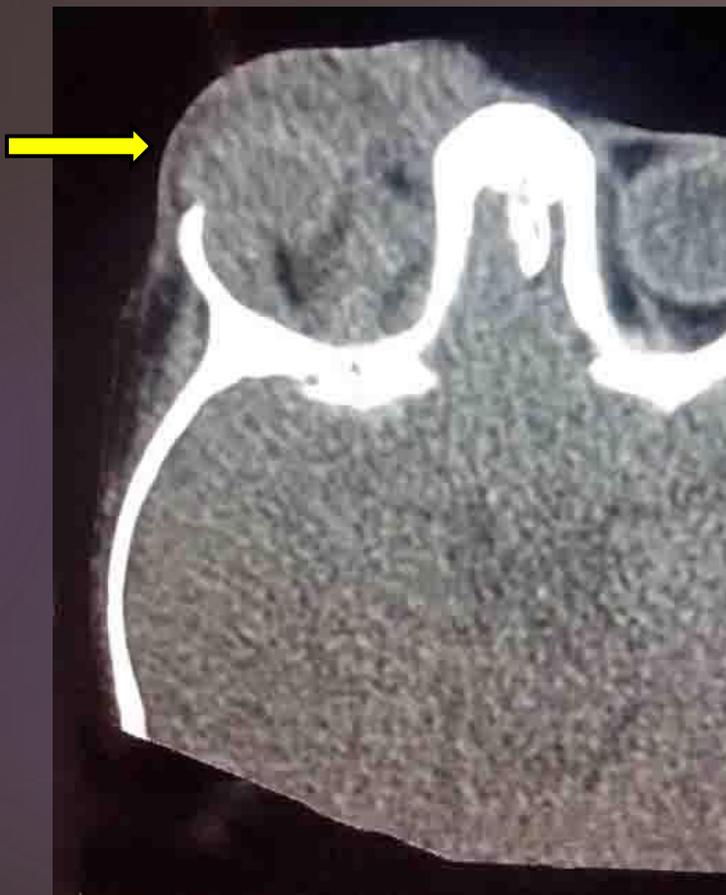
PADUA, PAULA FRANCESCA C.

OBJECTIVES

- ▶ To present a **case of venolymphatic malformation** & OK-432 as its management
- ▶ To discuss the **mechanism of action** of OK-432
- ▶ To discuss the known **complications** of OK-432



CT SCAN



Injection of OK-432



**Before
Therapy**



**1 Day
Post-Therapy**



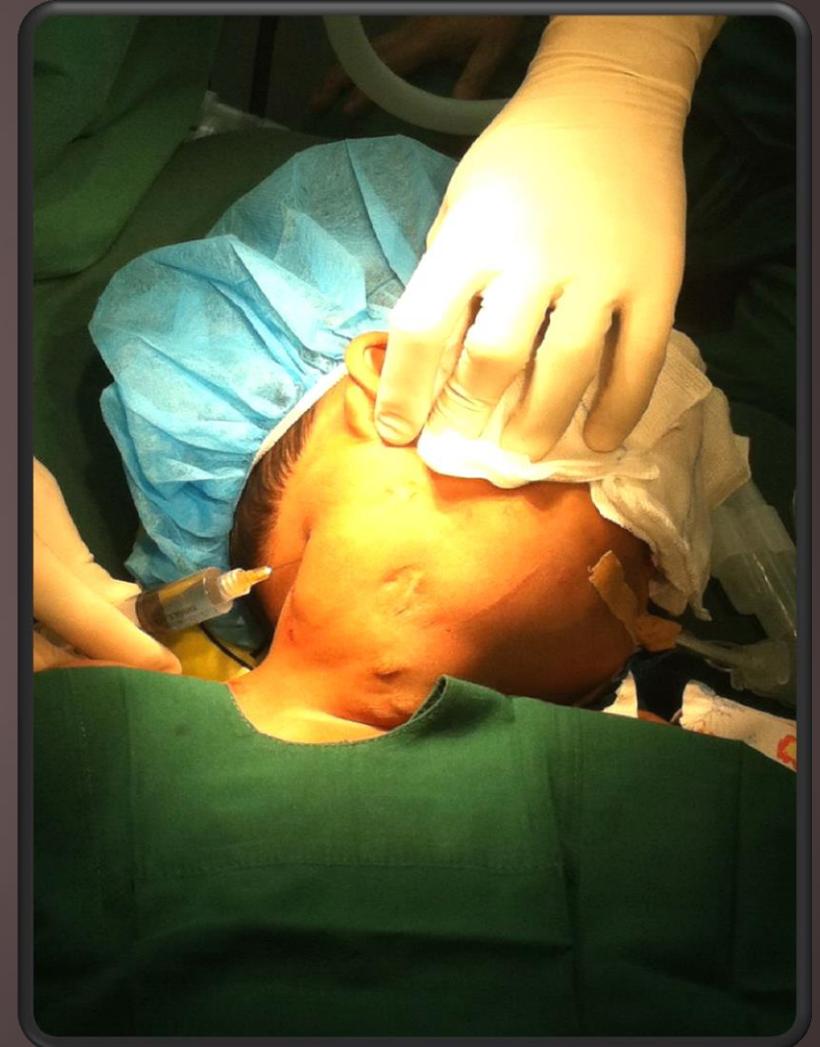
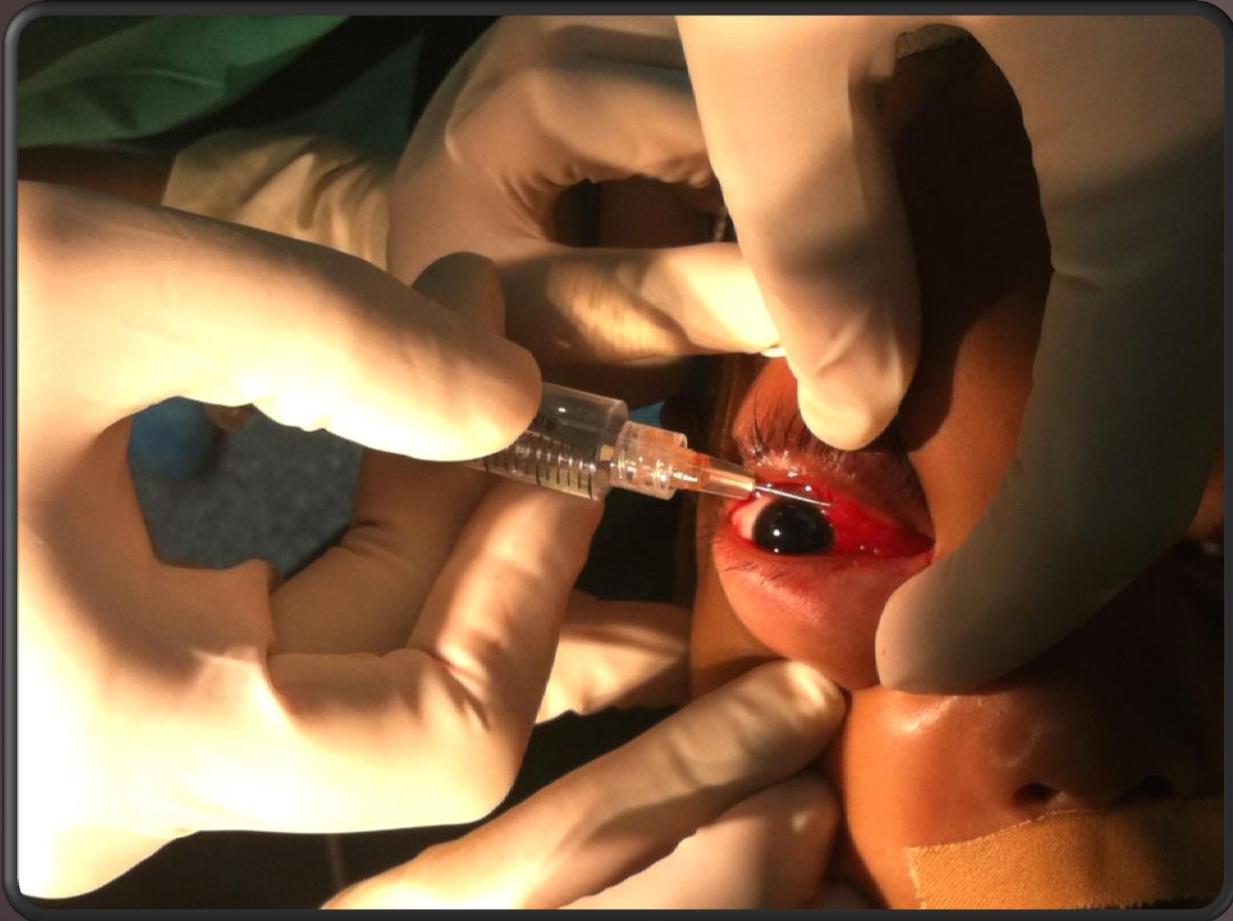
**2 Mos
Post-Therapy**



**3 Mos
Post-Therapy**



2nd Injection of OK-432





**Respiratory
distress**

**Hypotensive
episodes**

**Decreased
sensorium**

Sepsis?

Equivocal
Serum Pro-Calcitonin

Negative Cultures:

ETA

Urine

Blood

CSF

= Not Sepsis

Anaphylaxis?

Elevated

IgE

Hours after event:

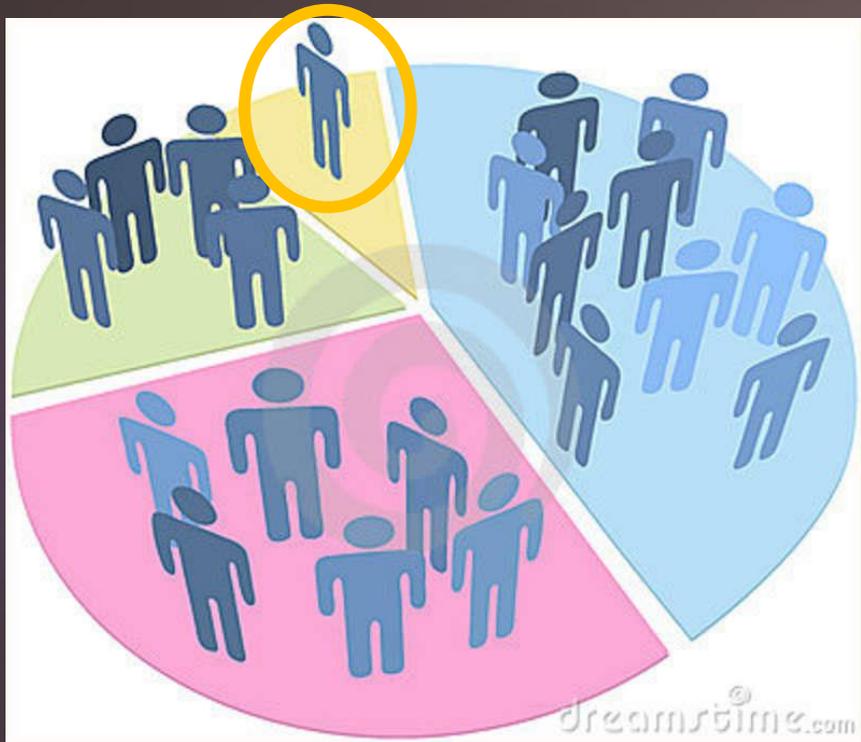
>24 Hours

= Not Anaphylaxis

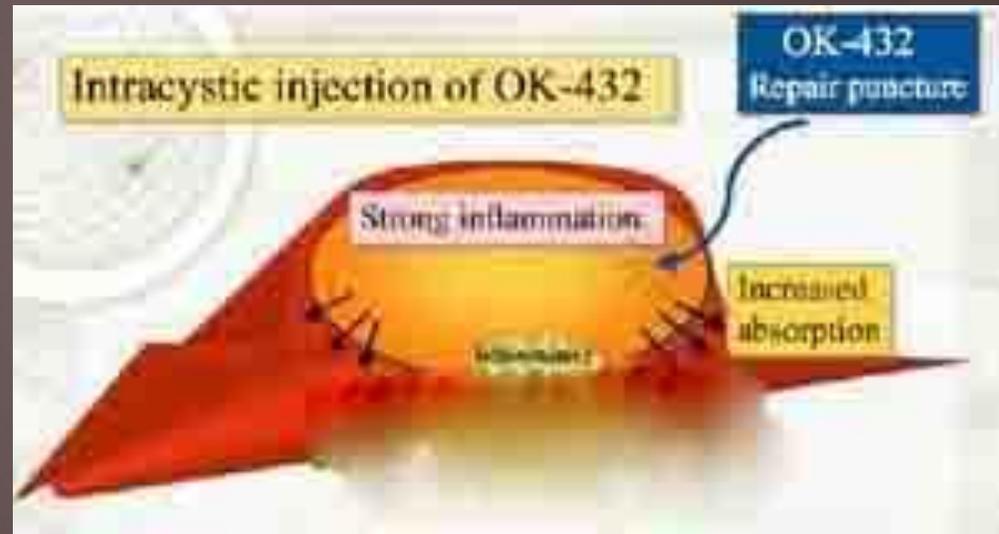


So what is it then?

Venolymphatic Malformation



OK-432 (PICIBANIL)



COMPLICATIONS

- ▶ Local inflammatory reaction
- ▶ Fever

SERIOUS SIDE EFFECTS

~~► Hypopharyngeal edema~~

The first serious side effect encountered with OK-432, which is soft tissue swelling after **Dexamethasone** hypopharyngeal edema with swallowing problems.

Laranne, et al

Intubation

...15% of all children reviewed required a tracheostomy tube.

Boardman, et al

SERIOUS SIDE EFFECTS

- ~~▶ Hypopharyngeal edema~~
- ~~▶ Systemic Immunomodulatory Response~~

Injections induce systemic immunomodulatory responses with sepsis-like, life threatening

Ultrasound Guidance

Check for backflow

this would

Bittmann, et al



SERIOUS SIDE EFFECTS

- ▶ ~~Hypopharyngeal edema~~
- ▶ ~~Systemic Immunomodulatory Response~~
- ▶ ~~Anaphylaxis~~

Other potential risks include hypersensitivity to either the bacteria-derived protein or to the penicillin component within the preparation (used for bacterial breakdown). That is why hypersensitivity to penicillin must be ruled out before treatment... These kinds of complications would all develop within a few hours after Picibanil injections.

Skin Test

Bittmann, et al

~~Lymphocytopenia...~~

▶ Another possible cause?

Patients with large abscesses often have low numbers of bacteria, making them at risk for bacterial dissemination secondary to reduced immunomodulatory response.

Procalcitonin = Equivocal, *et al*

Systemic Inflammatory Response Syndrome?

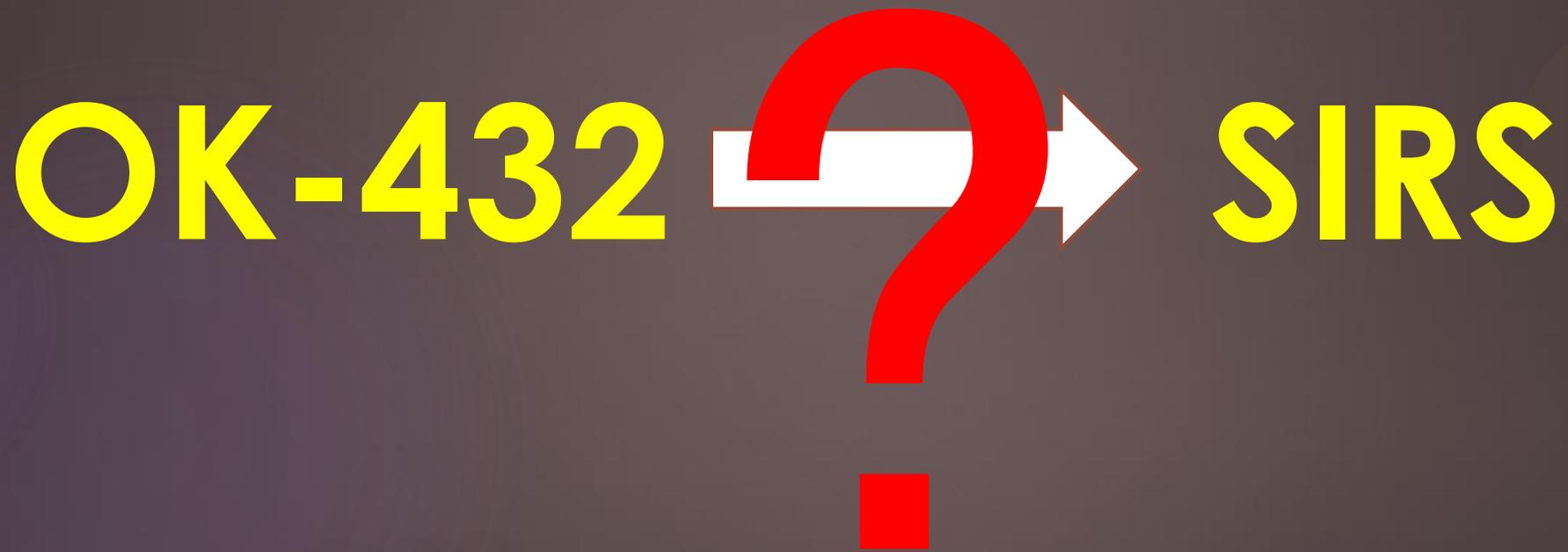
Table 1 Definitions associated with the SIRS (adapted from Bone *et al.*, 1992)

SIRS

The systemic inflammatory response to a variety of severe clinical insults. Manifested by two or more of the following:

- (1) body temperature $>38^{\circ}\text{C}$ or $<36^{\circ}\text{C}$,
- (2) heart rate >90 beats per minute,
- (3) tachypnoea: respiratory rate >20 breaths per minute or $\text{PaCO}_2 <4.3$ kPa (or 32 mmHg),
- (4) white blood cell count $>12\,000/\text{mm}^3$ or $<4000/\text{mm}^3$ or $>10\%$ immature bands.

These changes should represent an acute change from baseline and be unexplained by other causes.



Conclusion

- ▶ OK-432 showed promising results of regression of malformations in published literature.
- ▶ We are presented with a case of SIRS after OK-432 use.
- ▶ Further studies are recommended to establish if there is an association between SIRS and OK-432 use.

Thank You!