

# Developing a national program for care of patients with Retinoblastoma

Sidnei Epelman, M.D.

INCTR  
São Paulo  
Brasil

# Developing World Pediatric Oncology

- 80% - 160.000 newly diagnosed cases per year
- Access to care
- Human resources
- Essential medications
- No cancer registries
- High morbidity and mortality
- Lack of support

# Pediatric Oncology

Cure

Psychological support

Adequate therapeutical support

Adequate treatment

Precise diagnosis

Comprehensive center

Early diagnosis

# Retinoblastoma

- Affects newborns, infants and toddlers, and most children are diagnosed before 2 ½ years of age
- Each year there are an estimated 8.000 cases worldwide and of these cases, 7.000 children die from the disease due to the delay in diagnosis or to lack of access to expert medical care
- Higher incidence in less developed countries

# Retinoblastoma

- The management of retinoblastoma has gradually changed over the past 10 years
- Over 95% of children with retinoblastoma in the world could be cured with modern techniques.
- The challenge remains, however, in maintaining the eye and vision

# Retinoblastoma Program

- Education professional and lay population
- Establishment of local resources for diagnosis and treatment
- Universal health care access

# The Context

- Multidisciplinary Team
  - Pediatric Oncologist
  - Ophthalmologist
  - Radiation Oncologist
  - Supportive Care
- **Priorities in Treatment**
    - Cure
    - Eye Salvage
    - Vision Preservation

# Diagnosis

- Examination under Anesthesia
- Retinal Drawings
- Retinal Photographs
- RET-CAM
- Diagnostic Imaging

# Pathology of Retinoblastoma Staging

- Choroidal invasion
- Invasion of sclera
- Penetration of sclera along emissarial canals
- Extrascleral tumor around vessels
- Meningeal invasion
- Spinal fluid dissemination

# Treatment

- There is a trend away from enucleation and external beam radiotherapy toward focal conservative treatments involving primary chemoreduction in conjunction with thermotherapy and cryotherapy
- This is related to earlier detection of the disease, recognition of more effective chemotherapeutic agents, more focused local treatment modalities, and, most importantly, knowledge of the long-term risks of external beam radiotherapy

# Treatment

- Enucleation
- External beam radiation
- Chemotherapy
  - Radioactive implant
  - Cryotherapy
  - Laser therapy

# National Campaign of Retinoblastoma in Brazil



- More than 50% extra-ocular disease
- More than 8 months between first sign and diagnosis
- 400 cases per year
- Better treatment is not the solution
- Easy sign to educate lay population
- Campaign initiated in September 2002



EM 75% DOS CASOS  
DE TUMOR OCULAR,  
O BRILHO NO OLHAR VOLTA.

**T U C C A**

Associação para Crianças e Adolescentes  
com Tumor Cerebral  
[www.tucca.org.br](http://www.tucca.org.br)  
0800 770 4665.

 PATROCÍNIO 

**INCTR**  
**2007**



QUANTO ANTES O  
TUMOR OCULAR FOR DETECTADO,  
MAIS CHANCES TEM DE SER CURADO.  
CAMPANHA PARA DIAGNÓSTICO PRECOCE  
DO RETINOBLASTOMA.

Uma foto do seu filho pode revelar  
muitas coisas, inclusive o retinoblastoma.

O retinoblastoma é um tumor maligno, originado das células da retina. Sua apresentação mais comum é o reflexo pupilar branco ou "olho de galo". Este sinal pode ser notado em algumas posições do olhar sob a luz de lâmpadas caseiras, ou através de fotos tiradas por câmeras simples com flash. O mais importante é que, se for diagnosticado precocemente, o retinoblastoma é uma doença curável, inclusive preservando a visão da criança. Olhe nos olhos do seu filho. Eles podem estar pedindo a sua ajuda.



Associação para Crianças e Adolescentes  
com Tumor Cerebral

www.aacc.org.br 0800 770 4665



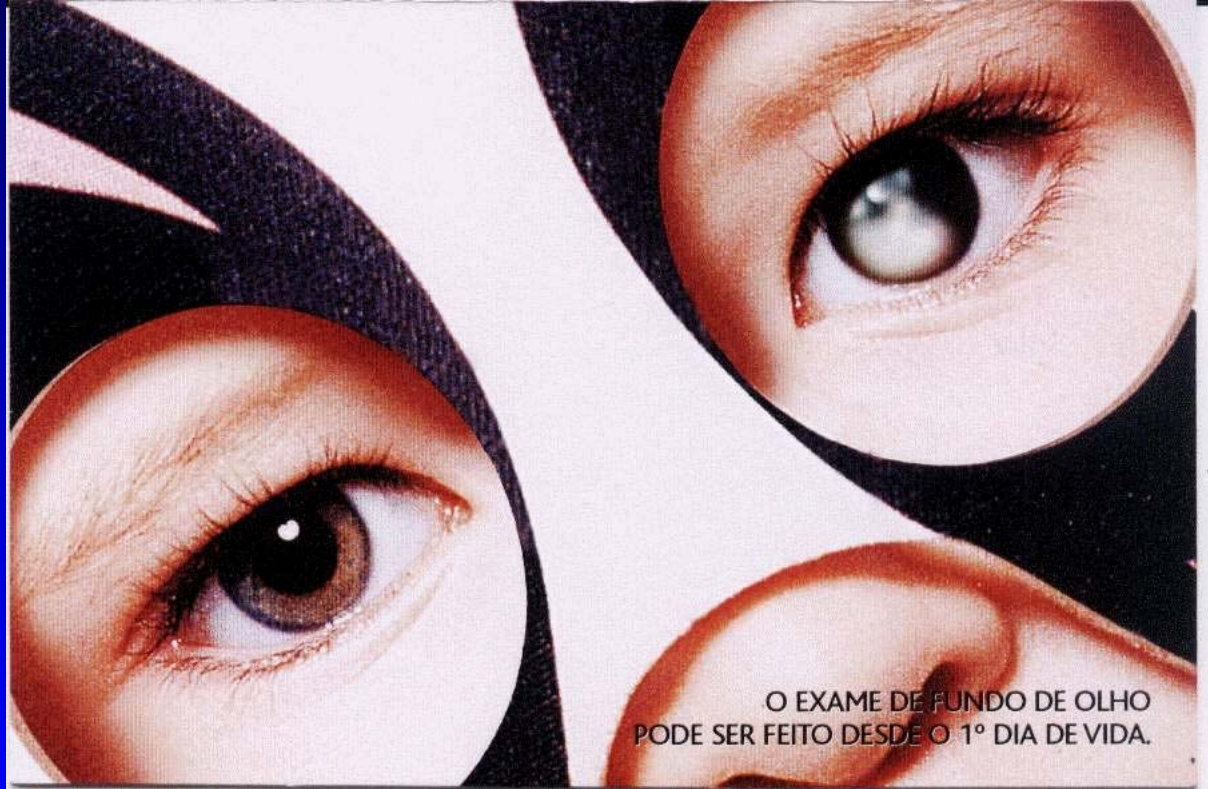
1983

Poster

INCTR  
2007



Ophthalmologist's card



## Pediatrician's card

**Quanto antes o tumor ocular for detectado,  
mais chances tem de ser curado.**

Campanha para diagnóstico precoce do retinoblastoma.



Associação para Crianças e Adolescentes  
com Tumor Cerebral

[www.tucca.org.br](http://www.tucca.org.br)

**0800 770 4665**



APOIO



# Actions

- Discussion with government to develop a law to perform fundoscopy in newborns in Brazil
- Translation of the video to 8 languages : english, french, arabic, spanish, hindu and has been distributed through International Network for Cancer and Treatment Research - INCTR
- Information to Maternities
- 2.500.000 phone cards for public telephones in the state of São Paulo
- Raise funds
- Toll free number - 0800 7704665



FOTOGRAFE SEU FILHO  
COM FLASH.  
EM CASO DE  
"OLHO DE GATO",  
CONSULTE UM MÉDICO.

Fale até  
80 min\* 40

**Quanto antes o tumor ocular  
for detectado,  
mais chances tem de ser curado.**

Campanha para diagnóstico precoce do retinoblastoma.

**TUCCA**

Associação para Crianças e Adolescentes  
com Tumor Cerebral

www.tucca.org.br 0800 770 4665

Apoio:

FUNDAÇÃO

Telefônica

"A mensagem publicitária contida neste cartão é única  
e exclusivamente de responsabilidade do anunciante."

\* ligação local  
para telefone fixo.

Interprint  
Tecnologia CPiD  
DNF: 029798-XXXX738

Tiragem = 200.000  
Fabricação: 06/2003  
Validade: 1 ano

27\*

- Não dobre nem amasse este cartão.
- Este cartão não é reembolsável.

**40**  
UNIDADES

Série: Tucca (04/04)

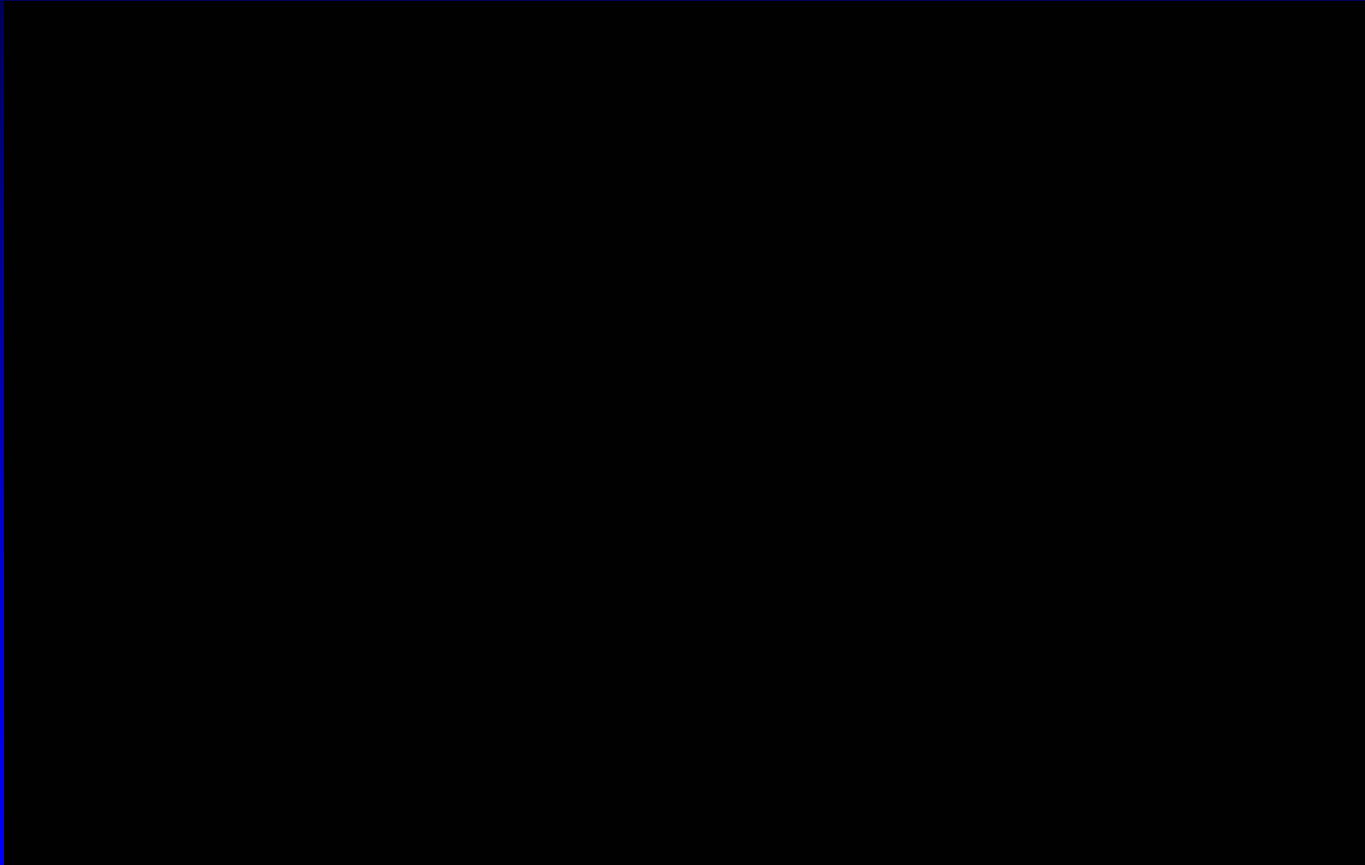
Informações sobre duração de  
cada unidade de crédito deste  
cartão, ligue 08007715104

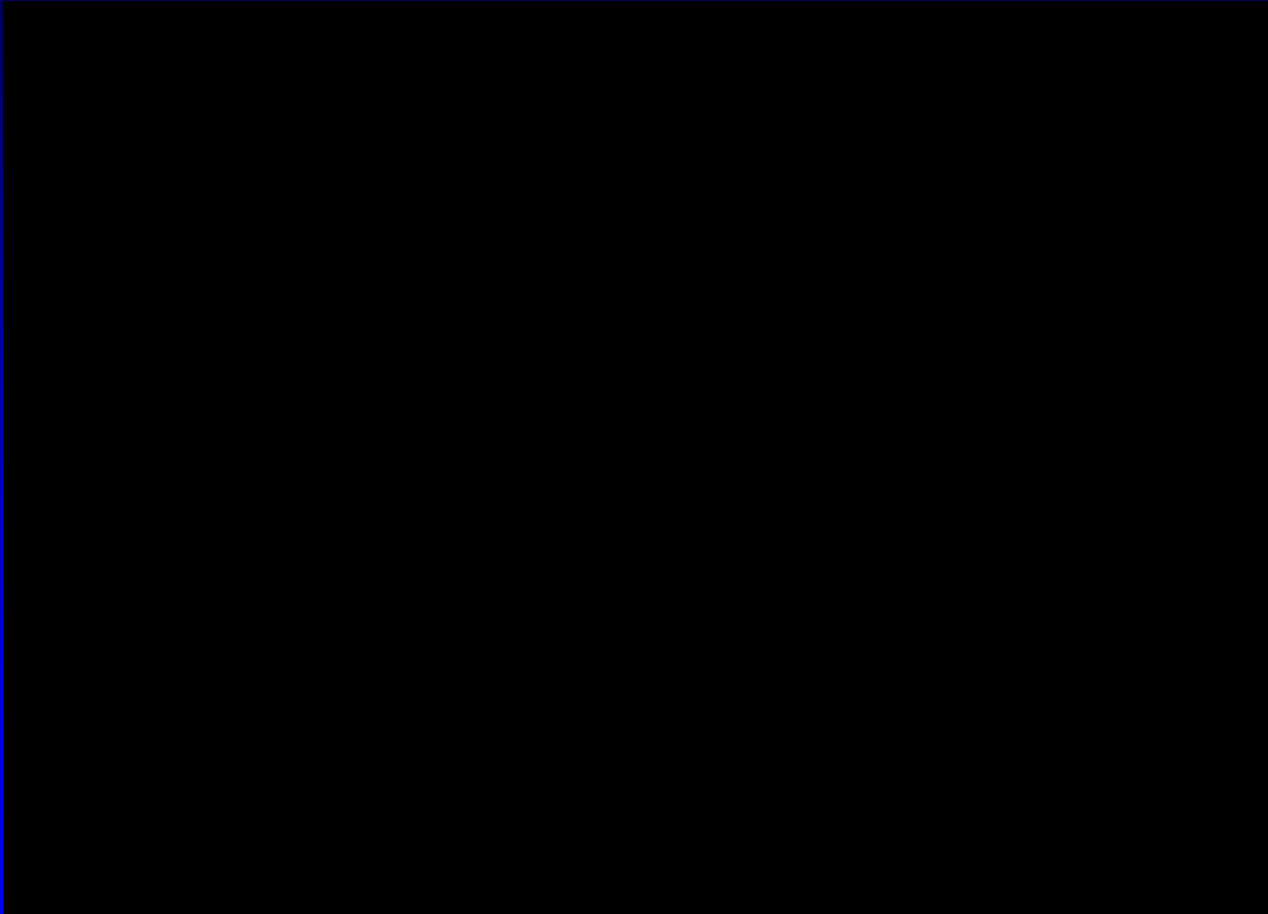
*Telefônica*



TEL 000042







# New actions

- Retinoblastoma Day
- Latin american meeting
- Qualify pediatricians and neonatologists to fundoscopy
- RetCam for screening

# Results

- Number of cases diagnosed through the campaign in all country - 50 (not all cases reported to us)
- Other ocular diseases diagnosed through the campaign: retinopathy of prematurity, familial exudative vitreoretinopathy, cataract, ocular toxocariasis, toxoplasmosis, Coat's disease
- Prevention of Blindness in childhood



3-years old - live 15 days by boat in Amazon Forest 2007



7-days old

# Screening

- Treatment or early detection must result in decreasing morbidity and mortality,
- Incidence in certain specific age must be high to justify the screening
- Risk population must be easily identifiable
- A natural history of the cancer must be well known
- The screening must be easily practicable and acceptable for the target population

# Screening for retinoblastoma

- Patients who had a family history of retinoblastoma and were clinically screened for retinal tumors from birth were diagnosed younger (8 months of age) and earlier (Reese Ellsworth group 1 = 26 [58%] of 45) and had better ocular survival than nonscreened patients with a family history. More patients were initially detected by family/friends (1315 [80%] of 1632) than pediatricians (123 [8%] of 1632) or ophthalmologists (156 [10%] of 1632).

# Screening

- Initial disease detection at the point of leukocoria or strabismus correlated with high patient survival rates and poor ocular survival rates for the presenting eye. Saving eyes and vision requires disease recognition before leukocoria, as demonstrated by the better ocular salvage rate among patients who had a positive family history and received clinical surveillance via early, routine dilated funduscopic examinations by an ophthalmologist

# California 1998

- 521.265 live births
- 70 retinoblastomas – diagnosis at 18 months
- 50/70 advanced disease at diagnosis
- 700 congenital cataracts – 70% missed first 3 months if screened
- 1/677 live births affected with an eye disease will cause blindness unless detected early
- 200-300 will be legally blind
- Cost to the state over lifetime - \$2 million/blind child

# Newborn screening program

- Phenylketonuria - 1/18000
- Maple syrup urine disease - 1/268000
- Galactosemia - 1/57000
- Biotinidase deficiency - 1/80000
- Retinoblastoma - 1/15000

# Screening Eyes' test

- Dilated pupil exam significantly enhances the detection of blinding and life-threatening birth defects arising inside the eye
- Guidelines for a dilated pupil exam by the primary care physician or during vaccination in early infancy facilitate detection and timely referral to an ophthalmologist for diagnosis and treatment

# Screening

- Pediatricians don't perform a dilated pupil exam as part of well-baby care when the child is discharged from the hospital because it is not included in the standard of care
- Early diagnosis and intervention can save millions of dollars annually
- Cost in time and supplies for the pupillary dilation test is negligible

# Retinoblastoma awareness day

- Raise awareness
- Increase understanding of the solution that exist
- Translate knowledge into action
- Generate a movement that stimulates collective responsibility and action