Pediatric Retinal Vascular Diseases

From Angiography to Vitrectomy

Ulrich Spandau
Sang Jin Kim

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Preface

Dear reader,
This book provides comprehensive and up-to-date information on diagnosis, medical, and surgical treatments for pediatric retinal vascular conditions, which are leading causes of childhood blindness throughout the world. Experienced ophthalmologists in this field discuss basic knowledge about these diseases, practical aspects of management such as exam under anesthesia, up-to-date diagnostic approaches including spectral-domain handheld optical coherence tomography (OCT), and OCT angiography. A high emphasis is placed on recent advances in medical and surgical treatments for pediatric retinal vascular diseases. Step-by-step instructions are given for the surgical treatment with anti-VEGF treatment, laser photocoagulation, and vitrectomy. Both the general ophthalmologist who cares for children with retinal diseases and the specialist (pediatric ophthalmologists and vitreoretinal surgeon) will find this book to be an informative resource in providing best care for children with pediatric retinal vascular conditions.

The book includes many videos, which demonstrate the surgeries step-by-step. All videos are listed in the Video list and can be accessed under http://extras.springer.com/Search. Enter the ISBN number of your book and download the videos.

Alternatively, the online version of every chapter contains the videos. Note the following footmark at the beginning of every chapter:

"**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-030-13701-4_16) contains supplementary material, which is available to authorized users." Copy and paste the https address in your browser and you can access the videos.

Uppsala, Sweden — Ulrich Spandau
Seoul, South Korea — Sang Jin Kim

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I want to thank my family and especially my wife Katrin for her never-ending patience with a husband who spends so much time with his books.

Ulrich Spandau

Contents

Abbreviations

FA	fluorescein angiography
FEVR	familial exudative vitreoretinopathy
GA	gestational age
LE	left eye
LIO	laser indirect ophthalmoscopy
PHPV	persistent hyperplastic primary vitreous
PMA	postmenstrual age
RE	right eye
ROP	retinopathy of prematurity

List of Videos

The videos can be accessed under http://extras.springer.com/Search. Enter the ISBN number of your book and download the videos.

Video 16.1	Injection of Lucentis for ROP
Video 21.1	Insertion of trocars
Video 21.2	Vitrectomy for ROP stage 4B RE
Video 21.3	Vitrectomy for ROP stage 4A LE
Video 21.4	Bilateral vitrectomy for ROP stage 4A and 4B (long audio)
Video 21.5	ROP stage 4B (very short)
Video 22.1	Encircling band and ROP
Video 23.1	ROP redetachment_short
Video 23.2	ROP 4A redetachment
Video 24.1	Stage 4B with fibrovascular membranes
Video 24.2	Tractional detachment secondary to fibrovascular membranes
Video 24.3	Tractional detachment secondary to fibrovascular membranes (short)
Video 24.4	Pediatric cataract with 27G
Video 27.1	Neurofibromatosis
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Video 27.3	PHPV with 27G
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Video 28.2	ROP with exudates fellow eye
Video 28.3	Bilateral vitrectomies for ROP stage 4A and 4B (short)
Video 28.4	ROP redetachment_short
Video 28.5	ROP 4A redetachment
Video 28.6	ROP stage 4B with exudates
Video 28.7	Bilateral ROP stage 4 A with exudates (long)
Video 28.8	Bilateral ROP stage 4 A with exudates (short)
Video 28.9	Tractional detachment secondary to fibrovascular membranes
Video 28.10	Removal of encircling band

Part I
Pediatric Retinal Diseases

Chapter 1
Coats Disease

1.1 Diagnosis of Coats Disease

1.1.1 Introduction

Coats disease is an idiopathic retinal vascular disorder characterized by retinal telangiectasia, exudation, and exudative retinal detachment. In 1908, George Coats first described case series with retinal telangiectasia and massive exudation [1]. Coats disease occurs most commonly in males in the first or second decades, but it can be diagnosed at any age. The majority of cases are unilateral, but recent studies using wide-field fluorescein angiography revealed that subclinical abnormalities such as peripheral nonperfusion are common in contralateral eyes [2, 3]. The clinical manifestations of Coats disease are highly variable, ranging from telangiectasia only to phthisis bulbi.

1.1.2 Pathogenesis

1.1.2.1 Histopathology

A histologic study on enucleated eyes with Coats disease revealed macrophage infiltration and cholesteric clefts in the subretinal space [4]. Retinal vascular abnormalities were also demonstrated including dilated vessels with hyalinized vessel walls [4]. Immunoreactivity for VEGF was observed in the detached retina, dilated vessel, and macrophages infiltrating the subretinal proliferative tissue [4]. VEGFR-2 immunoreactivity was also observed in endothelial cells located in abnormal retinal vessels and inner layer of the detached retina, but not in macrophages infiltrating the subretinal space [4].

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1.1.3 Genetics

Previous studies reported mutations in several genes including *NDP* [5], *CRB1* [6], *TINF2* [7], *PANK2* [8], and *ABCA4* [9] in patients with Coats disease or Coats-like retinal phenotype. However, the exact molecular mechanisms remain to be elucidated.

1.1.4 Clinical Characteristics

1.1.4.1 Fundus Findings [10–12]

Retinal vascular telangiectasis (Figs. 1.1 and 1.2) develop most commonly in the inferior and temporal quadrants between the equator and the ora serrata [12]. Affected vessels show irregular and aneurysmal dilations. Vascular leakage from

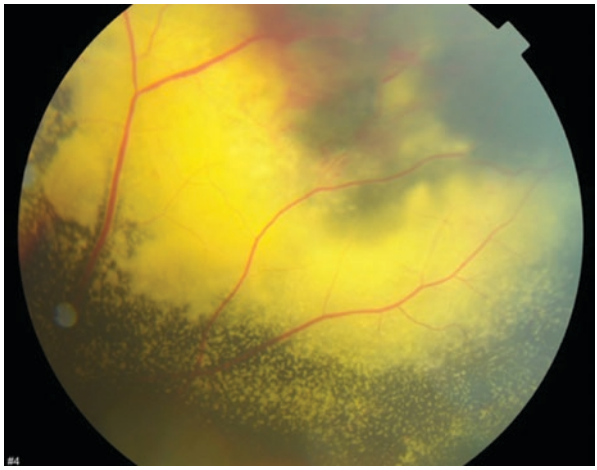

Fig. 1.1 Peripheral telangiectatic vessels with massive exudation and retinal detachment

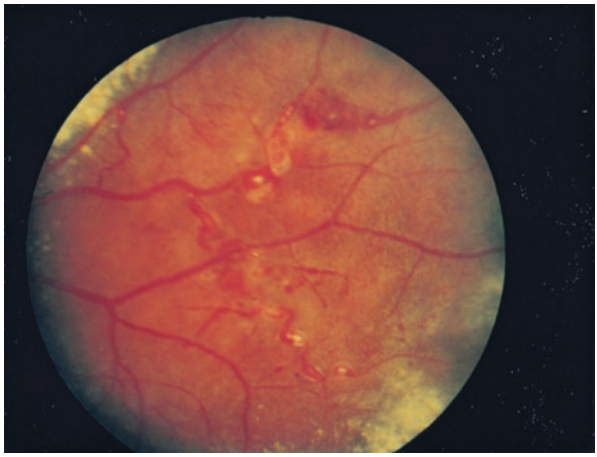

Fig. 1.2 Typical area of retinal telangiectasia without associated exudation. (Reprinted from Shields et al. [12]. Copyright (2001), with permission from Elsevier)

the abnormal vessels result in lipid-rich exudation (Figs. 1.3 and 1.4) and progressive fluid accumulation with subsequent serous retinal detachment (Figs. 1.5, 1.6, and 1.7) [11]. Macular edema or subretinal fluid is a common cause of visual symptom.

Retinal pigment epithelial cells that proliferate and migrate into the subretinal space may develop subretinal fibrous proliferation [11].

The vitreous usually remains clear [11]. Vitreoretinal traction, fibrosis, or proliferative vitreoretinopathy are not common but epiretinal membrane may develop [11].

In a large-scale case series (n = 150 patients) study by Shields et al. in 2001 [10], median age at the diagnosis was 5 years. Among the 150 patients, 114 (76%) were males and 142 (95%) showed unilateral involvement [10]. The most common referral diagnoses were Coats disease in 64 (41%) followed by retinoblastoma in 43 (27%) patients [10]. Visual acuity at presentation was 20/200 or worse in 121 eyes (76%) [10]. The retinal telangiectasia involved the midperipheral or peripheral

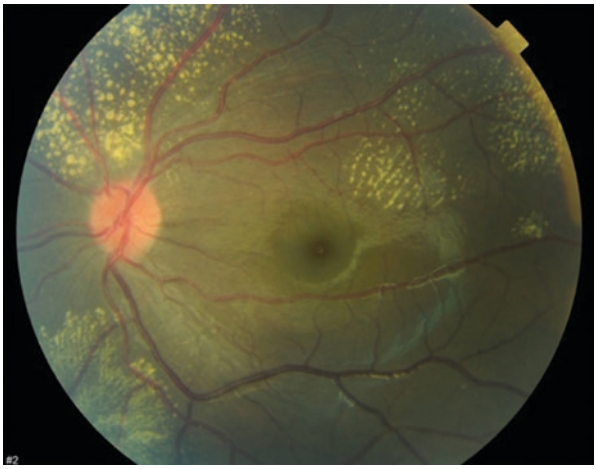

Fig. 1.3 Exudates at posterior pole in an 8-year old boy with Coats disease

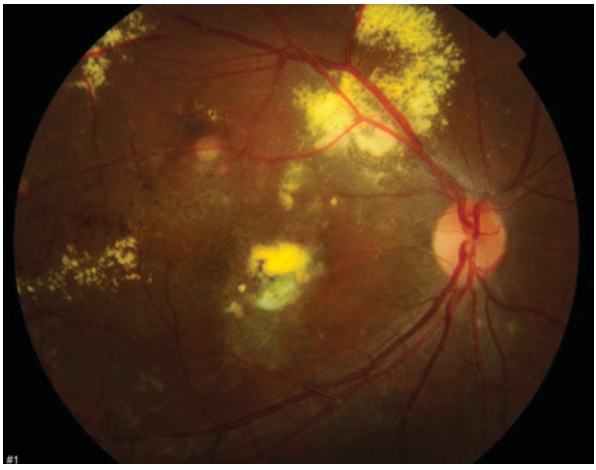

Fig. 1.4 Long-standing exudates at posterior pole in Coats disease

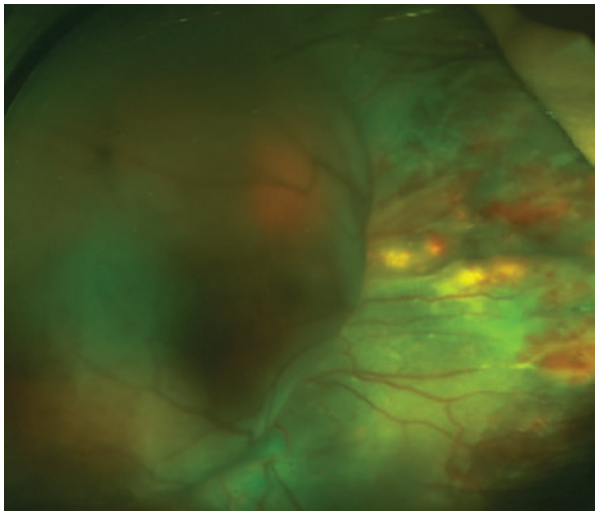

Fig. 1.5 Total retinal detachment in Coats disease

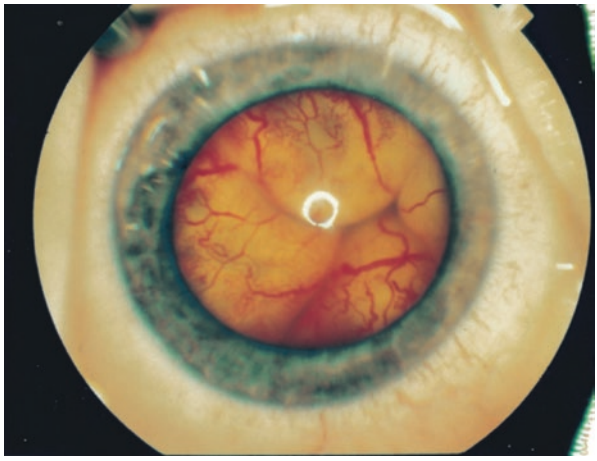

Fig. 1.6 Total retinal detachment in a patient with Coats disease. (Reprinted from Shields et al. [12]. Copyright (2001), with permission from Elsevier)

fundus in 98% of eyes [10]. Retinal exudation was present in six or more clock hours in 115 eyes (73%) [10]. Total retinal detachment was seen in 74 eyes (47%) and neovascular glaucoma in 12 eyes (8%) [10].

1.1.5 Fluorescein Angiography

Wide-field angiography systems such as RetCam (Natus) or Ultra-widefield™ retinal imaging systems (Optos) are essential in diagnosis and management of Coats disease. The angiographic features of Coats disease include areas of nonperfusion, peripheral telangiectatic capillaries and "light bulb" aneurysms, vascular leakage,