

Pediatric Retinal Vascular Diseases

From Angiography to Vitrectomy

Ulrich Spandau
Sang Jin Kim



Springer

EXTRAS ONLINE

Pediatric Retinal Vascular Diseases

Ulrich Spandau • Sang Jin Kim

Pediatric Retinal Vascular Diseases

From Angiography to Vitrectomy



Springer

Ulrich Spandau
Ophthalmology
University of Uppsala Ophthalmology
Uppsala
Sweden

Sang Jin Kim
Department of Ophthalmology
Samsung Medical Center
Sungkyunkwan University
Seoul
South Korea

Additional material to this book can be downloaded from <http://extras.springer.com>.

ISBN 978-3-030-13700-7 ISBN 978-3-030-13701-4 (eBook)
<https://doi.org/10.1007/978-3-030-13701-4>

Library of Congress Control Number: 2019935808

© Springer Nature Switzerland AG 2019

This work is subject to copyright. All rights are reserved by the Publisher, whether the whole or part of the material is concerned, specifically the rights of translation, reprinting, reuse of illustrations, recitation, broadcasting, reproduction on microfilms or in any other physical way, and transmission or information storage and retrieval, electronic adaptation, computer software, or by similar or dissimilar methodology now known or hereafter developed.

The use of general descriptive names, registered names, trademarks, service marks, etc. in this publication does not imply, even in the absence of a specific statement, that such names are exempt from the relevant protective laws and regulations and therefore free for general use.

The publisher, the authors, and the editors are safe to assume that the advice and information in this book are believed to be true and accurate at the date of publication. Neither the publisher nor the authors or the editors give a warranty, express or implied, with respect to the material contained herein or for any errors or omissions that may have been made. The publisher remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

This Springer imprint is published by the registered company Springer Nature Switzerland AG.
The registered company address is: Gewerbestrasse 11, 6330 Cham, Switzerland

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
Seoul, South Korea

Ulrich Spandau
Sang Jin Kim

Acknowledgments

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

Part I Pediatric Retinal Diseases

1	Coats Disease	3
1.1	Diagnosis of Coats Disease	3
1.1.1	Introduction	3
1.1.2	Pathogenesis.	3
1.1.3	Genetics	4
1.1.4	Clinical Characteristics	4
1.1.5	Fluorescein Angiography	6
1.1.6	Optical Coherence Tomography (OCT).	7
1.2	Classification of Coats Disease	9
1.2.1	A Classification System	9
1.2.2	Stage and Visual Outcome	10
	References.	13
2	Norrie Disease	15
2.1	Introduction	15
2.2	Genetics	15
2.3	Ocular Features	15
2.4	Extraocular Features	16
2.4.1	Auditory Findings	16
2.4.2	Neurological Findings	16
2.4.3	Peripheral Vascular Disease.	17
2.5	Management.	17
	References.	17
3	Incontinentia Pigmenti	19
3.1	Pathophysiology and Genetics of IP	19
3.2	Clinical Features	20
3.2.1	Ocular Manifestations	20
3.2.2	Retinal Screening Protocol	22

3.3	Skin Manifestations	24
3.4	Neurologic Manifestations	24
3.5	Other Manifestations	24
3.6	Diagnostic Criteria of IP	24
	References	25
4	Familial Exudative Vitreoretinopathy	27
4.1	Introduction	27
4.2	Pathophysiology and Genetics	27
4.3	Diagnosis of Familial Exudative Vitreoretinopathy	28
4.3.1	Clinical Features	28
4.3.2	Fluorescein Angiography Findings	30
4.3.3	OCT Features	31
4.4	Classification of Familial Exudative Vitreoretinopathy	33
	References	34
5	Retinopathy of Prematurity (ROP)	37
5.1	Pathophysiology of Retinopathy of Prematurity (ROP)	37
5.2	Classification of ROP	39
5.2.1	Classification System of Retinopathy of Prematurity (ROP)	39
5.2.2	Location and Extent of Disease	39
5.2.3	Stage of ROP	39
5.2.4	Plus Disease	42
5.2.5	Pre-plus Disease	44
5.2.6	Aggressive Posterior ROP (AP-ROP)	44
5.3	Screening Recommendations	46
	References	50
 Part II Examination		
6	Fundus Examination	53
6.1	Small Pupil	53
6.2	Indirect Binocular Ophthalmoscopy	53
6.2.1	Examination Technique	55
7	Wide-Field Fundus Photography	57
7.1	RetCam 3	58
7.2	PanoCam™	60
7.3	3nethra neo	60
7.4	Pictor	63
7.5	Icon	64
	Reference	66

8	Angiography of Newborn	67
8.1	Technique of Retcam Angiography	67
8.2	Retcam Angiography Atlas	68
8.2.1	ROP 3 in zone I	69
8.2.2	Incomplete Laser Photocoagulation for ROP Newborn.	69
8.2.3	ROP Stage 4.	70
8.2.4	ROP Stage 4B	72
8.2.5	Incontinentia Pigmenti.	72
8.2.6	Microcephalus	75
8.2.7	FEVR or ROP	75
8.3	Optos Angiography Pictures	76
8.3.1	Familial Exudative Vitreoretinopathy (FEVR)	76
8.3.2	Morbus Coats	76
	References.	78

Part III Assessment

9	Assessment of ROP Neonates	83
9.1	Assessment of Plus Disease.	83
9.2	Assessment Zone I or Zone II	86
	References.	90
10	Lasercoagulation or Anti-VEGF: What Is the Better Treatment?	91
	References.	93

Part IV Laser Photocoagulation

11	Technique of Lasercoagulation	97
11.1	Instruments for Laser Coagulation.	98
11.2	Laser Treatment Step-by-Step	102
11.3	In Conclusion.	107
	References.	107
12	Angiography Assisted Laser Photocoagulation for Newborn and Children	109
12.1	The Technique Step-by-Step	109
13	Inadequate Laser Coagulation	113
13.1	Case Series Report.	113

Part V Anti-VEGF Injection

14	Size of a Newborn Eye	119
	References.	121

15 Dose of Anti-VEGF Injection in Infants	123
15.1 Comparison of Intraocular Volume Between Infant Versus Adult Eyes	123
15.2 Dose of Intravitreal Bevacizumab (Avastin) for ROP	124
15.2.1 Dose of Bevacizumab in Case Series Studies	124
15.2.2 Results of a Phase 1 Dosing Study by Pediatric Eye Disease Investigator Group (PEDIG)	124
15.2.3 Practical Problems for Using Lower Dose of Bevacizumab.	126
15.3 Dose of Ranibizumab (Lucentis) for ROP.	126
15.3.1 Comparing Alternative Ranibizumab Dosages for Safety and Efficacy in Retinopathy of Prematurity (CARE-ROP) Study	126
References.	127
16 Technique of Anti-VEGF Injection	129
16.1 General Guidelines for Intravitreal Injection.	129
16.2 Considerations Before Injection	129
16.2.1 Clinical Setting	129
16.2.2 Bilateral Injections	130
16.2.3 Sedation.	130
16.2.4 Pre-existing Systemic/Ocular Conditions.	130
16.3 Intravitreal Injection Technique in Infants.	130
16.3.1 Pupil Dilation	130
16.3.2 Anesthesia.	131
16.3.3 Anti-sepsis.	131
16.3.4 Needle and Syringe.	131
16.3.5 Volume of Medication.	132
16.3.6 Location of Injection.	133
16.3.7 How to Inject at the Bedside.	133
16.3.8 How to Inject in the Operation Room with the Microscope	134
16.3.9 After Injection.	134
16.3.10 Follow-Up Examinations	134
References.	135

Part VI Failure, Recurrence and Follow-Up

17 Recurrence and Complications After Laser Coagulation and Anti-VEGF Treatment	139
17.1 Recurrence/Reactivation After Laser Coagulation.	142
17.2 Complication Rate for Laser Coagulation	142
References.	143
18 Combined Laser and Anti-VEGF Treatment for Zone I ROP	145
References.	147

19	Recurrence of ROP After Anti-VEGF Treatment	149
19.1	Introduction	149
19.2	Clinical Course Following Anti-VEGF Treatment	149
19.3	Incidence of Recurrence	150
19.4	Timing of Recurrence	150
	References.	152
20	Persistence of ROP Disease After Laser Coagulation or Anti-VEGF: What to Do?	153
20.1	Treatment Algorithm for ROP 3+ Disease for Treatment Centers and Non-treatment Centers	154
	References.	159
Part VII Surgery		
21	Lens-Sparing Vitrectomy (LSV) for ROP Stage 4A and B	163
21.1	Physiology of a Neonate Eye.	163
21.2	Retinal Detachment Secondary to ROP.	164
21.3	Timing of Surgery	165
21.4	Anatomical and Functional Outcome of Surgery for 4A and 4B Detachment	166
21.5	Surgery.	166
21.6	Complications	170
21.7	FAQ	170
21.8	Case Report No. 1: ROP Stage 4	170
	Further Reading	171
22	Scleral Buckling for ROP Stage 4A and 4B	173
	Reference	174
23	Posterior Hyaloid Contraction Syndrome	175
	Reference	176
24	Retinal Detachment Stage 4A and 4B with Fibrovascular Membranes	177
	Reference	181
25	Stage 5 ROP	183
	Further Reading	185
26	Visual Outcome of Very Preterm Neonates at 6.5 Years of Age	187
	References.	189

Part VIII Case Series Reports

27	Pediatric Retinal Diseases	193
27.1	Case Report No. 1: Neurofibromatosis Type 2	193
27.2	Case Report No. 2: Persistent Hyperplastic Primary Vitreous (PHPV)	194
27.3	Case Report No. 3: FEVR	196

27.4	Case Report No. 4: Congenital Familial Exudative Vitreoretinopathy (FEVR)	202
27.5	Case Report No. 5: Is this FEVR and ROP = ROPER?	206
27.6	Case Report No. 6: Incontinentia Pigmenti	206
27.7	Case Report No. 7: Morning Glory Syndrome	211
27.8	Case Report No. 8: Microcephalus	213
27.9	Case Report No. 9: Coats Disease	214
27.10	Case Report No. 10: Avastin and Laser Treatment for ROP Zone I.	216
27.11	Case Report No. 11: Lucentis Treatment for ROP Zone I.	219
27.12	Case Report No. 12: Three-Year Follow-Up After ROP in Zone I and Treatment with 1× Lucentis.	221
27.13	Case Report No. 13: Seven-Year Follow-Up After ROP in Zone I and Treatment with 1× Avastin.	225
27.14	Case Report No. 14: Seven-Year Follow-Up After ROP in Zone I and Treatment with 1× Avastin.	226
27.15	Case Report No. 15: Two-Years Follow-Up After ROP in Zone I and Treatment with 1× Lucentis.	226
27.16	Case Report No. 16: Retinal Bleedings 8 Years After Surgical Treatment for ROP.	227
27.17	Case Report No. 17: Delayed Treatment of ROP Plus Disease.	230
	References.	231
28	Treatment Failures of ROP Disease.	233
28.1	Case Report No. 18: 2× Recurrence After Laser Treatment for Zone I (Aggressive Posterior ROP)	233
28.2	Case Report No. 19: Vitrectomy for Stage 4A Retinal Detachment	235
28.3	Case Report No. 20: Bilateral Stage 4A and Stage 4B Detachment and Retinal Redetachment.	238
28.4	Case Report No. 21: Vitrectomy for ROP Stage 4b with Shallow Detachment and Exudates	242
28.5	Case Report No. 22: Laser Coagulation and Lucentis for Stage 4A Detachment After Inadequate Laser Treatment	244
28.6	Case Report No. 23: Delayed Treatment in a 22-Week Newborn.	247
28.7	Case Report No. 24: Intravitreal Lucentis for ROP Stage 4A After Cryo and Laser Treatment.	249
28.8	Case Report No. 25: Vitrectomy for ROP Stage 4A	251
28.9	Case Report No. 26: Failed Vitrectomy for ROP Stage 4B.	252
29	Interesting Case Reports in ROP from the Literature	257
29.1	Very Late Reactivation of ROP After Intravitreal Bevacizumab Injection.	257
29.2	Persistent ROP in Tetralogy of Fallot	258

29.3	Endophthalmitis After Intravitreal Injection for the Treatment of ROP	258
29.4	Exudative Retinal Detachment After Laser Photocoagulation or Anti-VEGF Injection in ROP	259
29.5	IOP Elevation After Intravitreal Anti-VEGF Injection	260
29.6	Other Unusual Responses After Intravitreal Anti-VEGF Injection in ROP	260
	References	261
	Index	263

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
Video 27.2	Intraoperative OCT
Video 27.3	PHPV with 27G
Video 27.4	Lens sparing vitrectomy for FEVR
Video 28.1	RE ROP stage 4A with exudates
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].

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

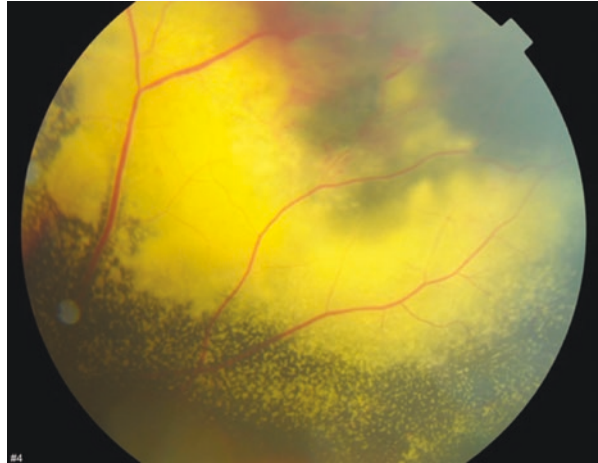
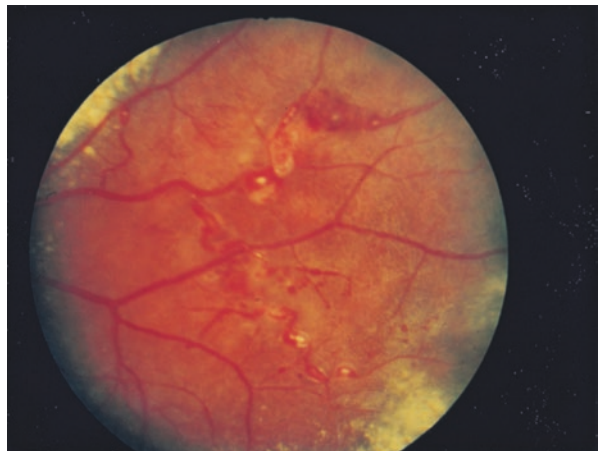


Fig. 1.2 Typical area of retinal telangiectasia without associated exudation. (Reprinted from Shields et al. [12]. Copyright (2001), with permission from Elsevier)



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

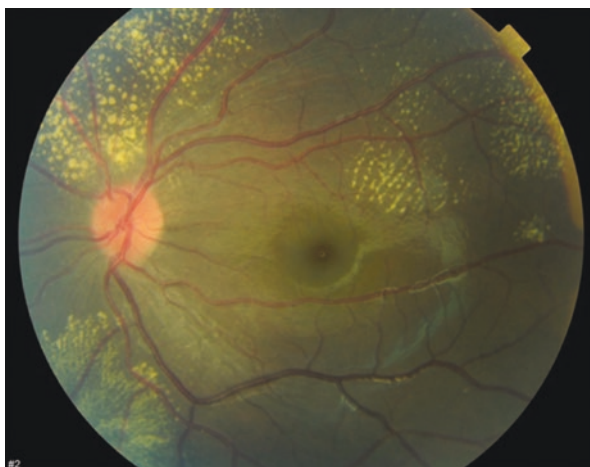


Fig. 1.4 Long-standing exudates at posterior pole in Coats disease

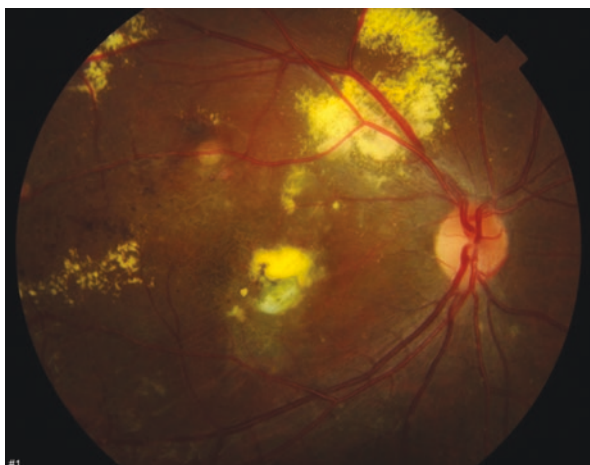


Fig. 1.5 Total retinal detachment in Coats disease

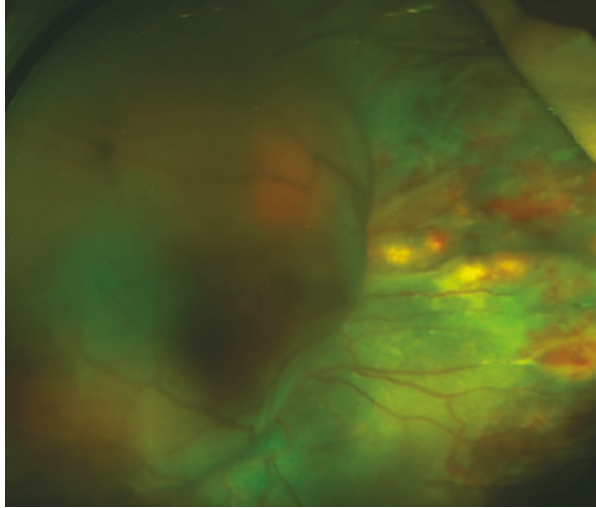
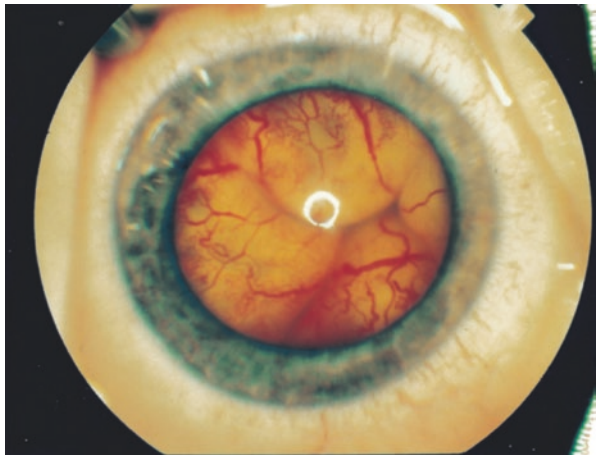


Fig. 1.6 Total retinal detachment in a patient with Coats disease. (Reprinted from Shields et al. [12]. Copyright (2001), with permission from Elsevier)



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,

Fig. 1.7 B-scan ultrasonography of total retinal detachment in a 12-year-old boy

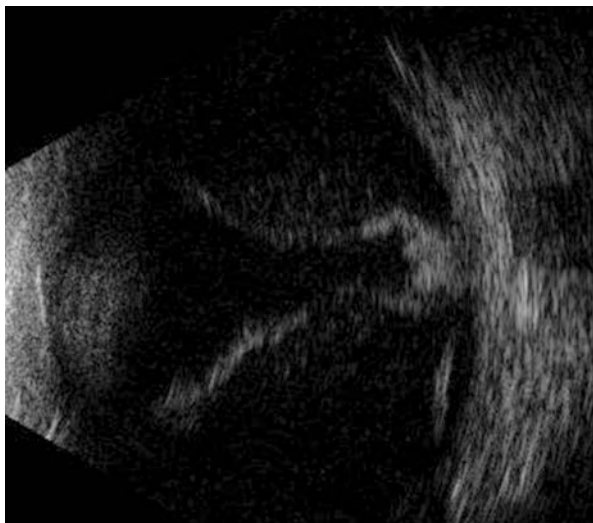


Fig. 1.8 Ultra-wide field fluorescein angiography showing dilated peripheral vessels with leakage in a patient with Coats disease



and blocked fluorescence from exudates (Figs. 1.8, 1.9, 1.10, and 1.11) [11]. Fluorescein angiography is essential in early detection of vascular abnormalities especially in eyes with telangiectasia only.

1.1.6 Optical Coherence Tomography (OCT)

In Coats disease, OCT is useful in identifying macular edema and subretinal fluid and to evaluate response to treatment. Subretinal fluid and exudate may be visible with OCT in patients with Coats disease (Figs. 1.12 and 1.13). It should be noted

Fig. 1.9 Ultra-wide field fluorescein angiography showing dilated peripheral vessels and nonperfusion in a patient with Coats disease



Fig. 1.10 Mild late leakage in normal-looking vessels in the inferior periphery in a patient with Coats disease



Fig. 1.11 Decreased leakage after cryotherapy in a patient with Coats disease



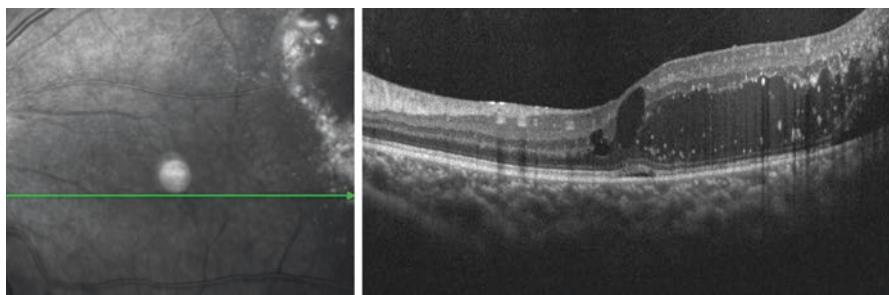


Fig. 1.12 SD-OCT showing macular edema and exudates in a patient with Coats disease

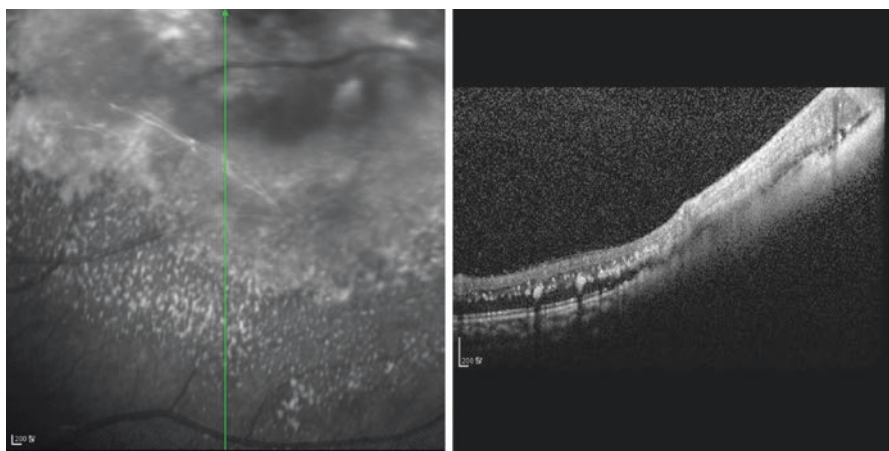


Fig. 1.13 SD-OCT showing intraretinal lipid deposits in a patient with Coats disease

that in eyes with large amount of subretinal fluid, the amount of subretinal fluid seen on OCT scans taken in a sitting position may be different from that in a supine position due to fluid shifting.

1.2 Classification of Coats Disease

1.2.1 A Classification System

Shields et al. proposed a classification system of Coats disease based on their clinical observations in 150 consecutive patients in 2001 [10]. Their proposed classification system is now being widely-used and very helpful for selecting treatment methods and predicting the visual outcomes (Table 1.1 and Fig. 1.14).

Table 1.1 Staging classification of Coats disease [10]

Stage	Findings
1	Retinal telangiectasia only
2	Telangiectasia and exudation
A	Extrafoveal exudation
B	Foveal exudation
3	Exudative retinal detachment
A	Subtotal detachment 1. Extrafoveal 2. Foveal
B	Total detachment
4	Total retinal detachment; glaucoma
5	Advanced end-stage disease

Eyes with stage 1 disease can be managed by either regular follow-up exams or laser photocoagulation [10]. In stage 1 disease, there is high probability that the eye can be salvaged and the visual prognosis is usually favorable [10]. However, stage 1 disease is rare in a real clinical practice probably due to no symptoms.

Eyes with stage 2 disease can be managed by laser photocoagulation or cryotherapy, depending on the extent and location of the disease [10]. In stage 2A, the visual prognosis is generally good [10]. Eyes with stage 2B are usually salvaged and the visual prognosis is fairly good [10]. Visual prognosis of eyes with a dense yellow gray nodule by the foveal exudation is usually worse [10].

Eyes with stage 3A disease can generally be managed by photocoagulation or cryotherapy [10]. Some of the patients with stage 3A1 disease (extrafoveal subtotal retinal detachment) in a sitting position can reveal subfoveal fluid in a supine position (thus stage 3A2). Even if the retinal detachment involves the fovea, it will resolve when the telangiectasias are treated [10]. Laser photocoagulation is less effective in areas of retinal detachment, and cryotherapy is often preferable in such instances [10]. However, Levinson and Hubbard reported good anatomical outcome of 577 nm yellow laser photocoagulation in 16 patients including 5 patients with stage 3B disease [13]. Patients with stage 3B with bullous detachment may require surgical treatment (e.g. external subretinal fluid drainage).

Patients who present with stage 4 disease are often best managed by enucleation to relieve the severe ocular pain [10]. Patients with stage 5 disease generally have a blind, but comfortable eye and require no aggressive treatment [10].

1.2.2 Stage and Visual Outcome

The staging system of Coats disease is helpful for selecting treatment and predicting the ocular and visual outcomes. In a case series study including 150 patients from 1975 to 1999, the visual outcome was generally poor [10]. The proportion of

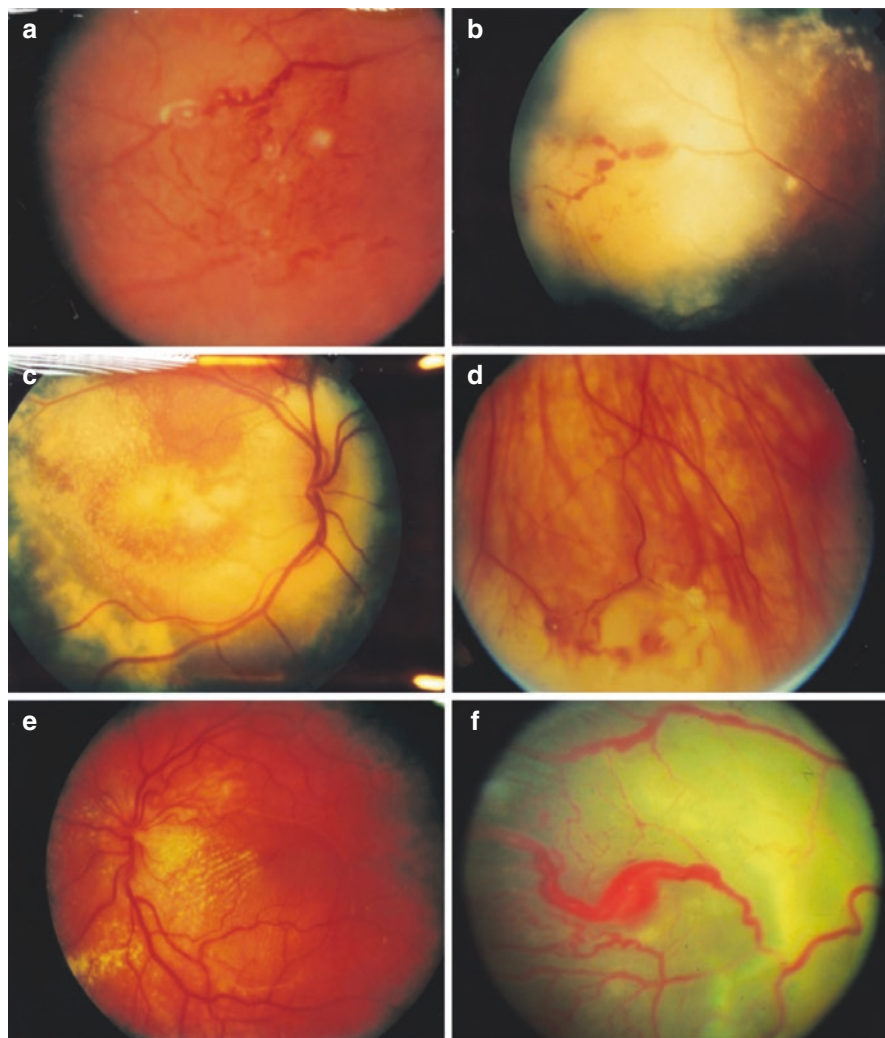


Fig. 1.14 Examples of stages of Coats disease. (a) Stage 1, retinal telangiectasia only. (b) Stage 2A, telangiectasia and extrafoveal exudation. (c) Stage 2B, foveal exudation. (d) Stage 3A1, subtotal retinal detachment inferiorly, sparing the fovea. (e) Stage 3A2, subtotal retinal detachment extending beneath the fovea. (f) Stage 3B, total exudative retinal detachment. (g) Stage 4, total exudative retinal detachment behind the lens in eye with secondary glaucoma. (h) Stage 5, advanced end stage disease with chronic inflammation, posterior synechia and cataract, secondary to longstanding retinal detachment. (Reprinted from Shields et al. [12]. Copyright (2001), with permission from Elsevier)

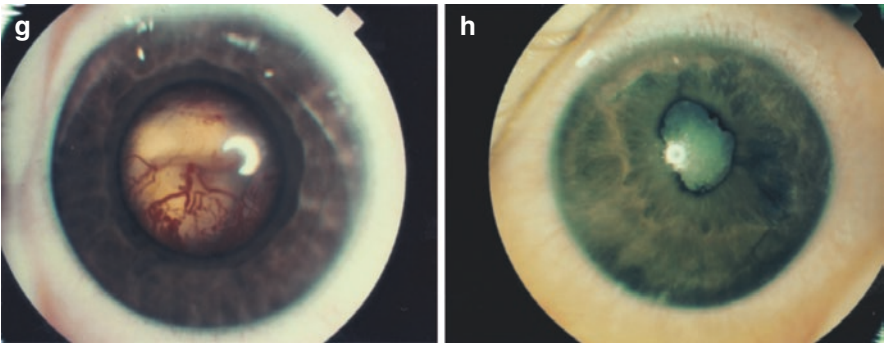


Fig. 1.14 (continued)

Table 1.2 Visual outcome according to the stage of Coats Disease

Stage	Shields et al. [10]		Ong et al. [14]			
	% poor visual outcome ^a	Number of eyes	1995–2005		2006–2015	
			% poor visual outcome ^a	Number of eyes	% poor visual outcome ^a	Number of eyes
1	0	1				
2A	30	10	50	2	0	1
2B	86	7	67	3	33	3
3A1	70	25	80	5	70	10
3A2	70	23				
3B	94	37	100	5	100	3
4	100	18	100	1		0
5	100	3		0		0

^aPoor visual outcome was defined as BCVA of 20/200 or worse

poor visual outcome (20/200 or worse) was high in eyes with stage 2B through 5 (Table 1.2). Recently, Ong et al. [14] compared visual outcome between two time periods (decade 1, 1995–2005 and decade 2, 2006–2015), and showed that (1) there was a trend for the mean initial presenting VA for decade 1 eyes to be worse than for decade 2 eyes; (2) from initial to final follow-up visit, mean VA also worsened for decade 1 eyes, but remained stable for decade 2 eyes; (3) at the end of follow-up, there was a trend for mean VA for decade 1 eyes to be worse than for decade 2 eyes; and (4) decade 2 eyes had a higher average number of procedures per eye compared with decade 1 eyes (Table 1.2). In conclusion, this study showed that the earlier presentation of disease in decade 2 suggests improvements in disease detection over time, and there was a trend for eyes to have better final VA in decade 2.