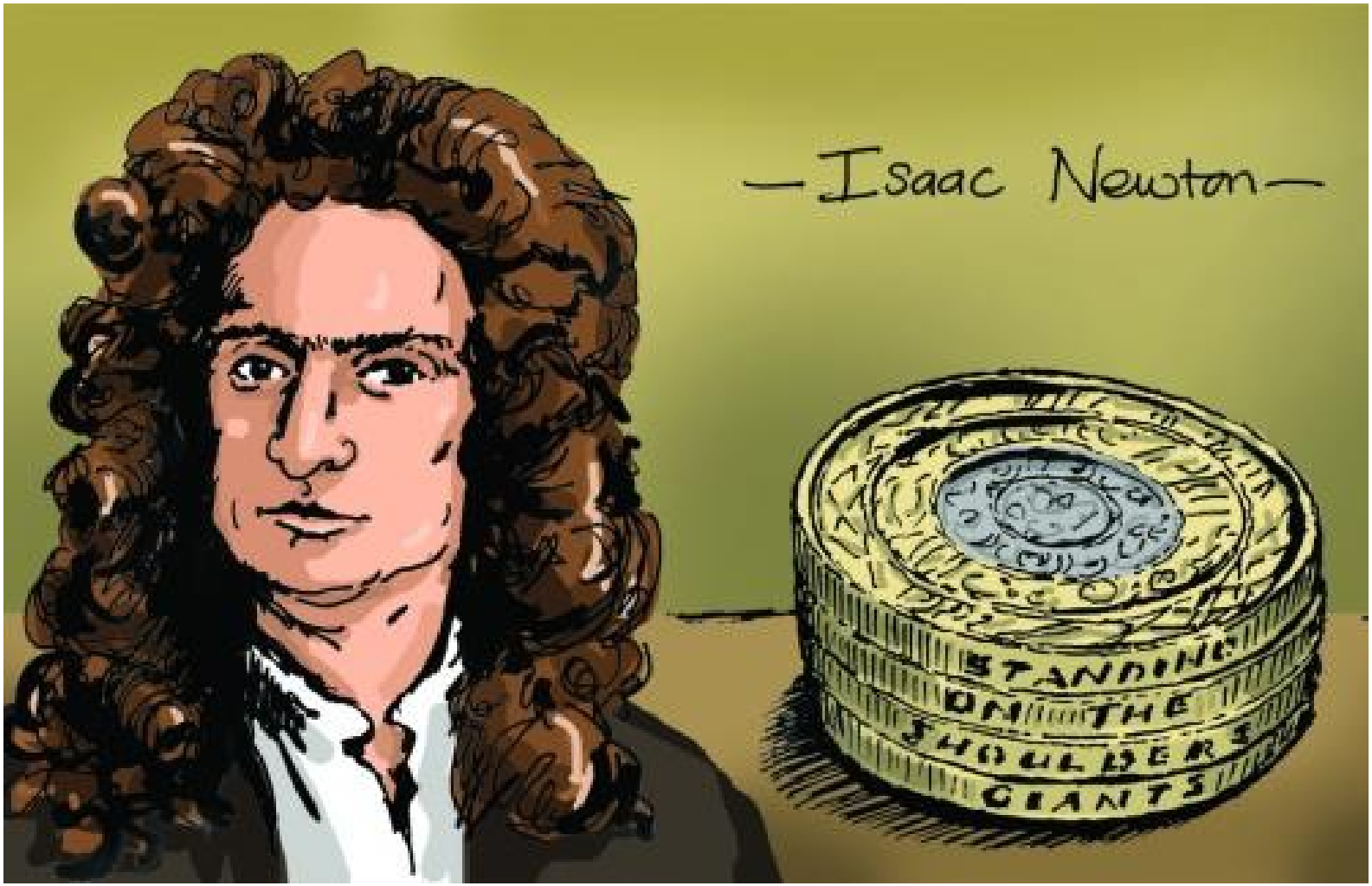
Save the Date

- 1st African Retinal Society Meeting
- Accra
- 23rd – 24th March
- Alisa Hotel
- Limited spaces

Surgical management of Proliferative Sickle Cell Retinopathy: A retrospective study of outcomes over the last two years.

Kwesi Nyan Amissah-Arthur
FRCOphth, FGCS
Senior Lecturer
University of Ghana Medical School

Ophthalmological Society of Ghana
September 2017


—Isaac Newton—
STANDING
ON THE
SHOULDERS
GIANTS

Incidence and Natural History of Proliferative Sickle Cell Retinopathy

Observations from a Cohort Study

Susan M. Downes, FRCOphth,[1,2] Ian R. Hambleton, PhD,[1] Elaine L. Chuang, MD,[1,3] Noemi Lois, MD,[1,2] Graham R. Serjeant, MD, FRCP,[1] Alan C. Bird, MD[1,2]

Objective: To describe the incidence, prevalence, and natural history of proliferative sickle cell retinopathy (PSR).

Design: Prospective longitudinal study over 20 years.

Participants: Newborn screening of 100 000 consecutive deliveries from 1973 to 1981 identified 315 children with homozygous sickle cell (SS) disease and 201 with SS–hemoglobin C (SC) disease. By the age of 5 years, 307 SS patients and 166 SC patients were alive and living in Jamaica and were recruited for this ophthalmic study.

Methods: Description of retinal vascular changes on annual angiography and angioscopy.

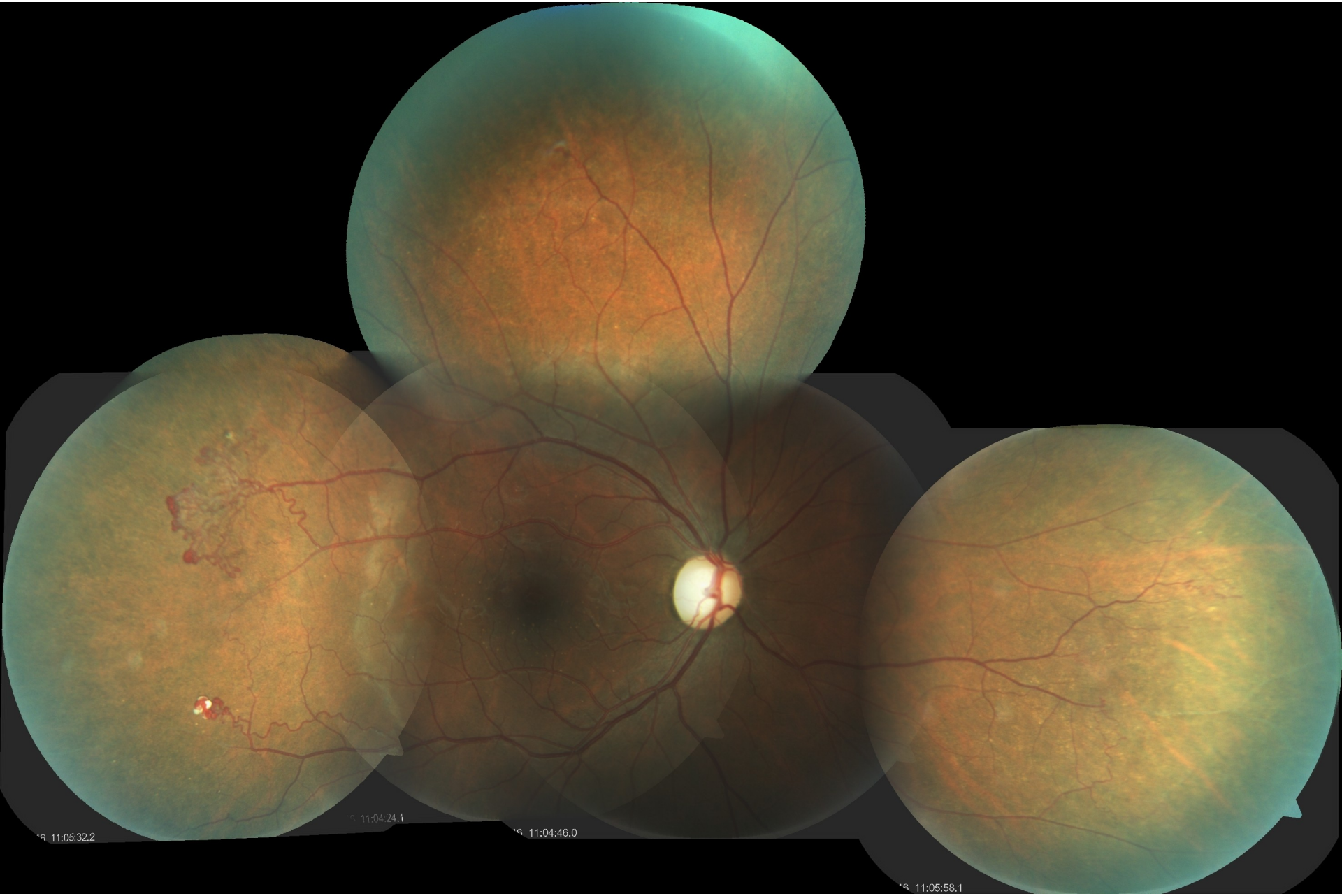
Main Outcome Measures: Incidence and prevalence of PSR and its behavior on follow-up. Progression of PSR was investigated using the number of eyes affected (none, one, both) and the interval until PSR onset.

Results: At last review in January 2000, PSR had developed in 59 patients (14 SS, 45 SC), unilaterally in 36 patients and bilaterally in 23. Incidence increased with age in both genotypes, with crude annual incidence rates of 0.5 cases (95% confidence interval [CI], 0.3–0.8) per 100 SS subjects and 2.5 cases (95% CI, 1.9–3.3) per 100 SC subjects. Prevalence was greater in SC disease, and by the ages of 24 to 26 years, PSR had occurred in 43% subjects with SC disease and in 14% subjects with SS disease. Patients with unilateral PSR had a 16% (11% SS, 17% SC) probability of regressing to no PSR and a 14% (16% SS, 13% SC) probability of progressing to bilateral PSR. Those with bilateral PSR had an 8% (8% SS, 8% SC) probability of regressing to unilateral PSR and a 1% (0 SS, 2% SC) probability of regressing to a PSR-free state. Irretrievable visual loss occurred in only 1 of 82 PSR-affected eyes, and 1 required detachment surgery and recovered normal visual acuity.

Conclusions: Longitudinal observations over 20 years in a cohort of patients followed from birth confirms a greater incidence and severity of PSR in SC disease, and shows that spontaneous regression occurred in 32% of PSR-affected eyes. Permanent visual loss was uncommon in subjects observed up to the age of 26 years.
Ophthalmology 2005;112:1869–1875

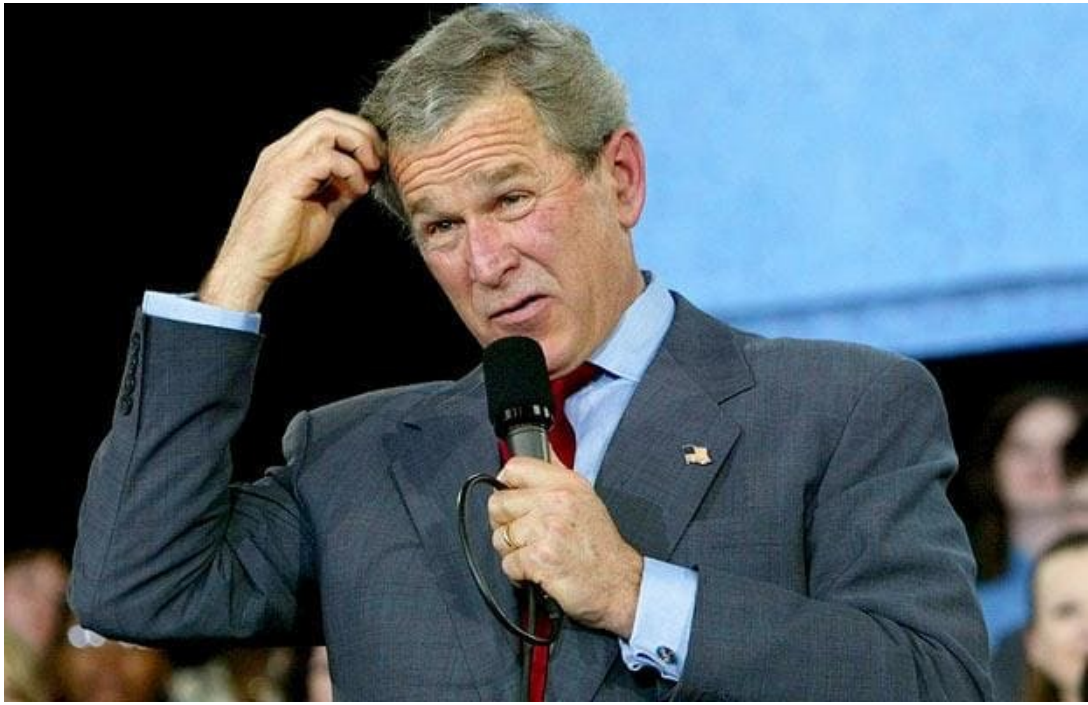
Challenging preconceived ideas

- 50 year old clergyman
- Not known Sickle Cell
- Non clearing Vitreous Haemorrhage
  - Flat on USSB
- HbSC diagnosed
- See in one month
  - Detachment!


11:05:32.2
11:04:24.1
11:04:46.0
11:05:58.1

Vitrectomy for Sickle Cell Retinopathy

- Books??
- Papers??
- Experience



Largest Operative Case series

18 cases

1996 - 2007

Prior to sutureless small gauge vitrectomy

CLINICAL STUDY

Vitreoretinal management of the complications of sickle cell retinopathy by observation or pars plana vitrectomy

TH Williamson, R Rajput, DAH Laidlaw and B Mokete

Department of Ophthalmology, St Thomas' Hospital, London, UK

Correspondence:
TH Williamson,
Department of Ophthalmology,
St Thomas' Hospital,
Lambeth Palace Road,
London SE1 7EH, UK
Tel: þ 0207 188 4320;
Fax: þ 0207 922 8157.
E-mail: tom@retinasurgery.co.uk

Received: 4 April 2008
Accepted in revised form: 12 August 2008
Published online: 3 October 2008

Financial Disclosures: None

Funding Support: None

Abstract

Purpose To describe the management of vitreoretinal complications of sickle cell retinopathy.
Design A retrospective interventional case series
Methods Review of an electronic patient record and clinical notes of 27 patients with vitreoretinal complications of sickle retinopathy.
Results Six male patients and 21 female patients presented with a mean age of 41 years (range 28–67), 12 left eyes and 16 right eyes. The mean follow-up was 15.5 months (range: 3–60). Two patients were SS, and the remainders were doubly heterozygous (SC). In all, 10 were observed without surgeryF three with vitreous haemorrhage, four tractional retinal detachments (TRD), and three rhegmatogenous retinal detachments (RRD). Two patients demonstrated spontaneous flattening of the retinaF one RRD and one TRD. Eighteen eyes had pars plana vitrectomies (PPVs)F seven with vitreous haemorrhage, three RRD, three TRD, three ERM (one bilateral), and two macular holes. In all, 15 patients (83%) had improved vision postoperatively. The mean logMar preoperative visual acuity was 1.07 (Snellen equivalent 6/70), SD 0.62 was significantly improved postoperatively (mean 0.42 (6/15), SD 0.48, P ¼0.001).
Conclusions Sickle retinopathy occasionally presents with vitreoretinal complications. These can often be observed and may spontaneously regress. If surgery is required eyes respond to vitrectomy procedures with segmentation of sea fan proliferation.

Eye (2009) 23, 1314–1320; doi:10.1038/eye.2008.296; published online 3 October 2008

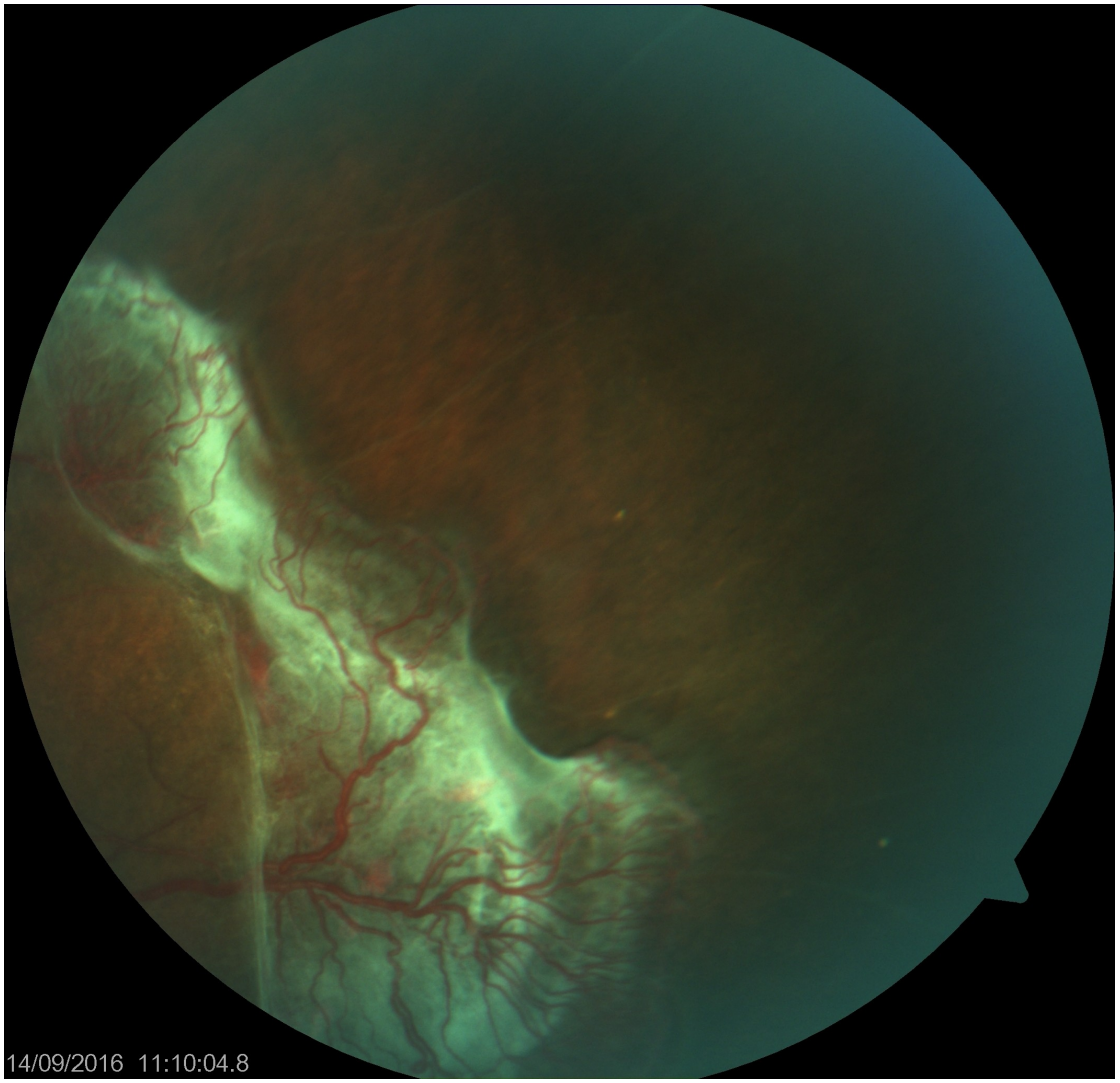
Keywords: sickle cell; retinopathy; vitrectomy

Introduction

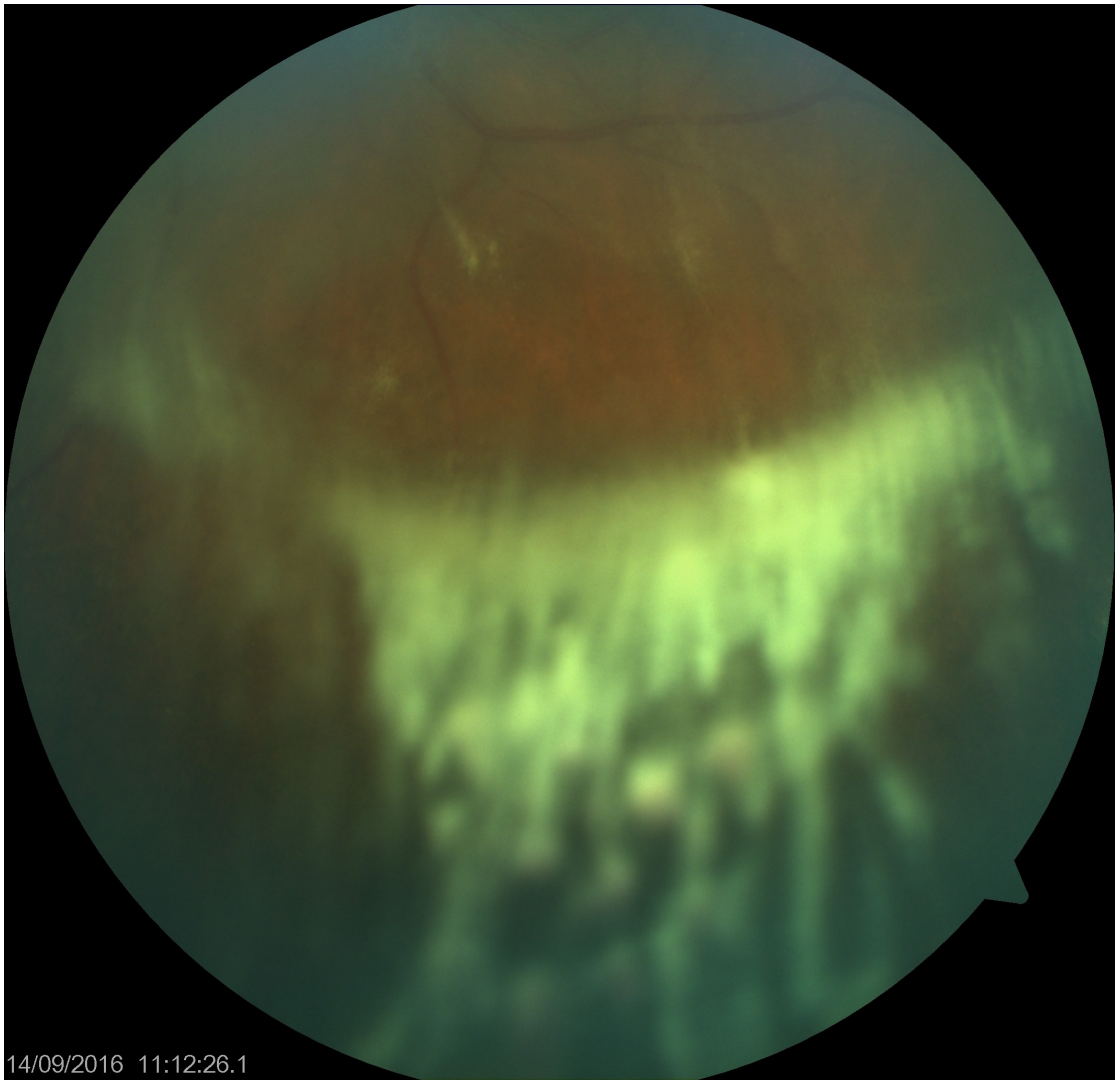
Sickle cell haemoglobinopathies result from an abnormality in the b-chain of the haemoglobin molecule. This causes chronic haemolytic anaemia and vaso-occlusive crises and a number of clinical features in the eye.[1,2] The conjunctival and optic nerve head blood vessels show characteristic segmentation of blood columns in the homozygous sickle cell disease (SS). In the retina, occlusion of the peripheral retinal vasculature occurs, accompanied by black sunburst spots, iridescent spots, retinal haemorrhages (salmon patches), and sea fan neovascular proliferation. Retinal complications are seen in 43% of patients aged between 20 and 30 years with doubly heterozygous (SC) sickle cell.[3] These include vitreous haemorrhage, tractional retinal detachment (TRD), rhegmatogenous retinal detachment (RRD), macular epiretinal membrane (ERM), and macular hole.[4] Reports of vitreoretinal surgery for these complications are few and tend to be from a period when scleral buckling and panretinal photocoagulation were used.[5–7] More recent reports on modern vitreoretinal techniques in sickle cell retinopathy are lacking and yet there has been considerable development of surgical technique. In this report, we reviewed the records of patients with vitreoretinal complications of sickle cell

Williamson, T. H., et al (2008). Vitreoretinal management of the complications of sickle cell retinopathy by observation or pars plana vitrectomy. *Eye*

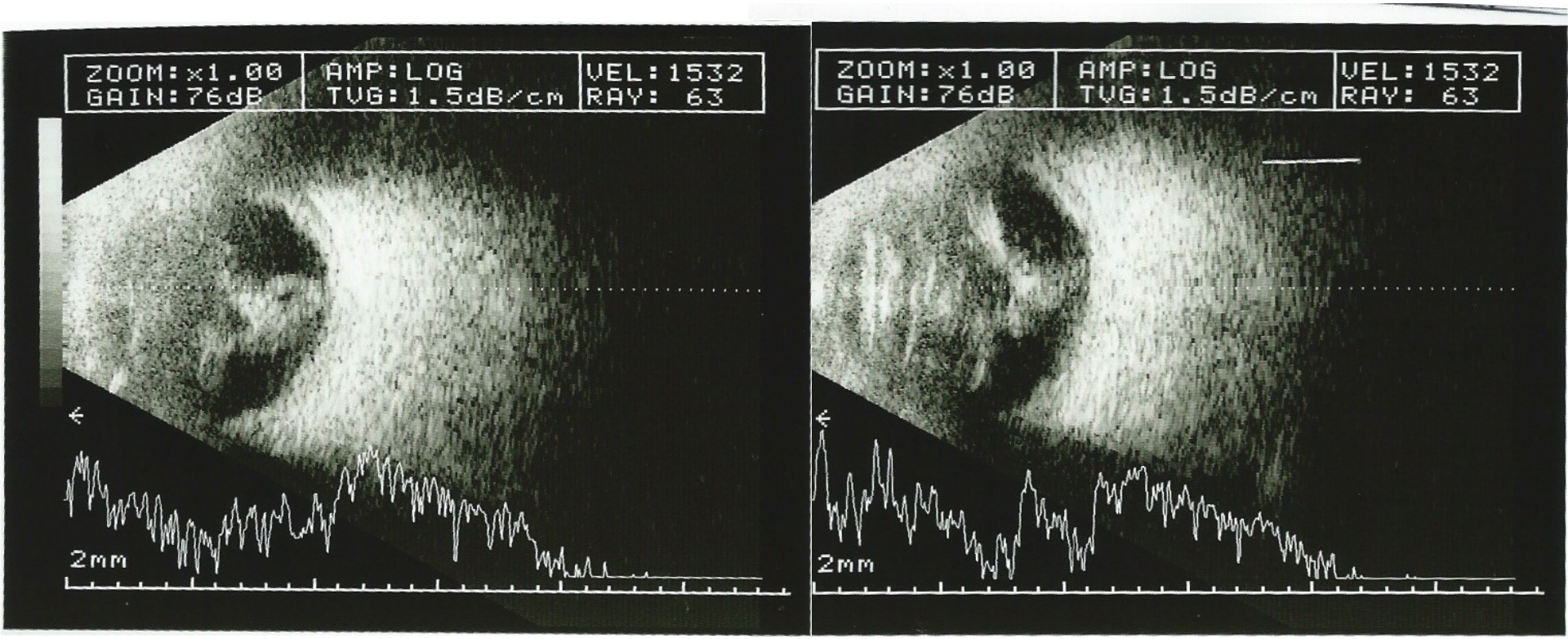
Stage 3 – Dr’s Akafo and Braimah



Stage 4 – Dr Seneadza, et al



Stage 5 – Dr Okonkwo et al



Last year OSG

- Advanced disease
  - Marked PVR
  - 2 cases – in one surgery abandoned
- Tractional retinal detachment
  - If minimal PVR and older age group appear to do well
- Macular Hole
- Non-clearing vitreous haemorrhage
  - Most satisfying

Research questions for VH & RD

- Do these patients require surgery or can we wait?
- What are the outcomes of surgery?
- Which surgical procedure is preferred?

Retrospective study

- Consecutive Interventional
- Minimum 6 months follow up
- Single Surgeon
- Operations at two hospitals

Presenting Demographics

- 18 cases met criteria
- Age: 26 – 48
- 3 Right and 15 Left
- Sex: F:6 M:12
- Presenting Va: 0.94 – 2.4(HM)
- **5 only eyes**
- Four Stage 4 and 14 stage 5

Indications for surgery

- Non Clearing Vitreous Hemorrhage: Four
- Retinal Detachment: 12
  - TRD: 10
  - Combined: 2
- Macular Hole: 2

Surgery

- Two Cryo-Buckle for Temporal limited RD
- Others were all 23G sutureless vitrectomy
- Three required re-operations
- Four required Silicone Oil

Outcomes

- Visual acuity ranged from 0.24 to NPL
- Mean VA 0.7 LogMAR
- 16 out of 18 patients had improvement in VA
  - 5 patients were better than 0.3 (6/12)
  - 13 patients were better than 1.0 (6/60)
- Had to give up on one patient intra-operatively
- Another patient had post-operative anterior segment ischaemia
- Two still have silicone oil insitu

Comparison to Williamson et al

Williamson et al

- 4 - RD
- 15/18 had improvement in VA
- One with SiOil
- *Fewer complications*

Our study

- 14 - RD
- 16/18 had improvement in VA
- Four with SiOil
- Two still insitu

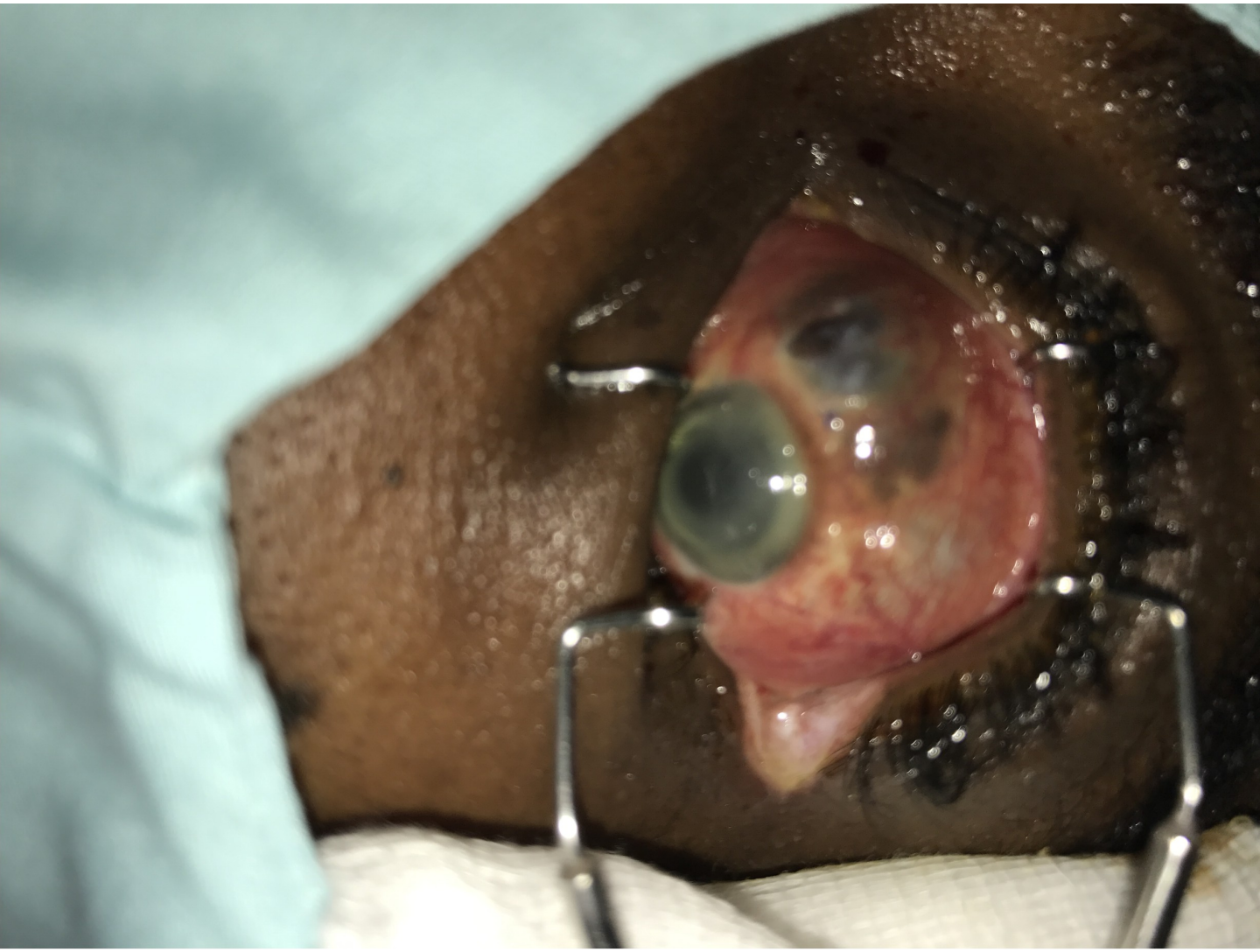
Limitations

- Retrospective study
- Limited follow up
- Surgeons bias

- **Payment bias**
- **Time to presentation bias**

Discussion

- Good improvement of vision in all VH Cases
- Retinal detachment cases can be complex
  - Younger cases still have attached Hyaloid
- Marked retinal scarring
- Buckle if possible
- High risk complications
  - Anterior segment ischaemia
- Abandoning a case on the table
- Patient selection



Thank You

Save the Date

- 1st African Retinal Society Meeting
- Accra
- 23rd – 24th March
- Alisa Hotel
- Limited spaces