Purpose: To present a case report of Coats disease and its management. Setting: Ophthalmology Department of Centro Hospitalar do Porto in Porto, Portugal. Methods: Patient’s data including clinical ophthalmologic evaluations complemented with medical and surgical procedures documented with retinographies and optical coherence tomographies. Results: A 6 year-old boy, previously healthy, was sent to our Department after being noticed a leukocoria in the left eye in a routine observation. The ophthalmologic evaluation revealed best corrected visual acuity of 10/10 (Snellen) in the right eye (RE) and hand movements in the left eye (LE) with a relative afferent pupillary defect in the last eye. Biomicroscopy of anterior segment was normal in both eyes. A mydriatic fundus examination was performed: in the RE was normal and in the LE showed retinal telangiectasia associated with a total exudative retinal detachment. An ecographic evaluation was done to rule out any intraocular tumor, which was negative. After discussing the therapeutic possibilities, it was decided to do surgery. The main procedures were: 1) two radial sclerotomies in the inferior quadrants deep enough to reach the suprachoroidal space to obtain the subretinal fluid drainage and 2) intravitreal injection of triamcinolone. One month later the retina was reattached and then underwent peripheral ablation. At the moment, the retina is still reattached although with no significant visual improvement. Conclusions: Coats disease is a rare disease that requires an early diagnosis in order to prevent its devastating complications. Vitreoretinal surgery can help to minimize consequences of the disease.