Good morning everyone, my name is Dupa and today I’d like to tell you about a 3-years-old boy’s case, but also a little bit about his illness in general because in low-resource management countries it remains a fatal disease.

Let me start by outlining a little about his condition. Peter was admitted to the hospital presenting chief complaints of redness, watering and white reflex in the right eye, persistent for the last 2 months. The left eye was within normal limits. He doesn’t have any past medical history, no history of trauma or eye infection. During his last visit to a GP, he was prescribed some eyedrops (local steroid, antibiotic and lubricant) but watering and redness were only improved. Regarding family history, his 15-years-old sister is diagnosed with thalassemia major, but there is no history of similar complaints in the family. Peter was born full-term, healthy and achieved his developmental milestones adequate for his age. He lives with his parents and his sister in a single-family house in Poznań. His mother doesn’t report any allergies.

Let me stop here for a moment because I would like to develop this point a bit further. Most of you probably catch the big red flag syndrome which is unilateral leukocoria also known as “white pupil syndrome”, so you already presume the diagnosis. What you must remember are other common clinical features typical for the disease such as poorly aligned eyes (professionally called strabismus), red and painful eye (usually due to glaucoma) or poor vision. Rarely the illness might be asymptomatic for a very long time.

Getting back to the little Peter’s case and his physical (or in this situation ocular) examination. Face, lid and eyelashes were normal in both eyes, ocular position and ocular movement of both eyes were full and free in all directions. Direct ophthalmoscopy showed right eye white reflex and sluggish pupillary reaction to light, the patient was not following the light. Sclera and iris detail were normal in both eyes. Afterward, the USG B-scan was performed which showed two large retinochoroidal mass lesions noted with the following dimensions with the specks of calcification seen posteriorly in the right eye. Next, the doctors carried out the MRI exam which reported a hypointense lesion in the right globe filling the vitreous in T2 and isointense signal in T1. No extraocular extension. This suggested retinoblastoma of the right eye with no abnormality in brain parenchyma. The diagnosis was confirmed by a histopathology report, which also showed no invasion of an optic nerve.

To sum up the diagnosis process- Ultrasonography is useful in distinguishing retinoblastomas from non-neoplastic conditions. It is also useful in detecting calcifications. MRI may be beneficial in estimating the degree of differentiation of retinoblastomas. Studies show that on T1-weighted images, the tumors usually have a low intensity and are usually difficult to distinguish from surrounding vitreous, but, on T2-weighted images, retinoblastoma tumors demonstrate very low intensity compared to vitreous. To be 100% correct, eye-cancer specialists would have to perform a biopsy. Blood counts and electrolyte determination as well as urinalysis and liver function tests are useful in excluding other conditions confused with retinoblastoma, which was not relevant in this case due to pathognomonic white-pupil syndrome. Normally differential diagnosis takes into account congenital cataract, retinopathy, toxocariasis, retinal dysplasia or retinal astrocytoma.

Before we move on now to the question of treatment, I’d like to quickly show you the international classification of retinoblastoma. There are five groups of advancement and medical therapy is individualized to the specific patient based on it. Peter was classified into group D and transported to a higher center for further management, where was performed enucleation with an 18mm optic nerve stump, followed by silicon ball implantation. Surgical removal of the tumor has been the standard management of very unfavorable retinoblastoma cases. The important thing is the younger the patient is at the time of surgery, the more growth retardation occurs. Post-op Peter was prescribed Amoxycillin and reviewed after 2 days. He also was given 1st circle of high-dose chemotherapy (1st out of 6).

What we have to realize is the prognosis in general is bad and the 5-year- survival depends on the extraocular extension. Fortunately, in Peter’s case after 1, 3, 6, 7 and 8 months since enucleation, there were no recurrence, no distance metastasis, no choroid and optic nerve invasion. He is now under medical supervision and turns 11 this year. And let me finish there. Thank you.