

# **CASE STUDY 2**

**“Examination of the Presentation  
and Treatment of a Patient with  
Suspected Retinitis Pigmentosa”**

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## **Introduction**

This study examines the case of a 12-year-old female who presented to the emergency department with her 10 year old sister and father. Consultation revealed a family history of retinitis pigmentosa (RP) and the subject was seeking confirmation or exclusion of the disease. RP is the most common retinal dystrophy and the most common inherited cause of blindness in people aged 20 to 60 years old. (Freeman, 1998:1)

This study will outline the actual and potential clinical signs and symptoms of RP, diagnostic methods employed and possible differential diagnosis. Further it will discuss the management, including current therapy and research being performed, and pharmacological information. It will also explore expected outcomes and possible complications of the disease. The subject of legal and ethical considerations is also discussed, particularly in relation to the subject being a child.

On a broader level the study aims to examine the epidemiology of RP throughout the world and within Australia. It will also outline the pathophysiological development of the disease. Included in the case study is a nursing care plan, which outlines areas for consideration when caring for such a patient.

## **Social History**

The subject lived with her parents and one younger sibling. She attended school and performed the activities of daily living of a 12 year old girl. Their mother had sent the girls to the department for assessment as she was “afraid they might have what their father had”. Questioning established that their father had been diagnosed with retinitis pigmentosa (RP) many years ago and was severely visually disabled as evidenced by his presentation being led by his daughter and using a cane for mobility.

## **Visual History**

The subject had worn glasses since the age of six for high myopia. When read the glasses were (R)  $-9.25/+2.75 \times 111$  and (L)  $-10.25/+3.75 \times 71$ . The subject reported onset of floaters in the right eye over the last two months. She also stated it had become more difficult to read the board at school over this period despite sitting at the front of the room. The subject reported a decrease in vision in low light conditions where she would “bump” into things at night.

## **Medical History**

Medical history revealed that the girl was generally well although she had suffered with pneumonia once in the past. No regular medications were taken. Family history revealed that the subject’s mother was diabetic. The father and paternal grandmother were both diagnosed with RP, the father having the poorer vision of the two. The subject’s sister, who had also presented for assessment, was a high myope.

## **Epidemiology**

The incidence of RP is relatively common and reported rates range between one in 3500 and one in 8000 births. (Chelva et al, 1992:311)

The prevalence of RP in communities of people varies. When recognizable syndromes are excluded the prevalence of RP in the following communities according to Weleber in Ryan (1994:336) were as follows:

<b>Community</b>	<b>Prevalence</b>
Maine (USA)	1 in 5200
Switzerland	1 in 7000
China	1 in 4016
Israel	1 in 4500
Navajo Indians	1 in 1878

In Australia a study of reported cases of RP from June 1976 to September 1988 cited a diagnosis of RP established in 707 patients from 461 different families examined at the Royal Victorian Eye and Ear Hospital, the Association for the Blind's Low Vision Clinic and private practices. (Chelva et al, 1992:313)

## **Pathophysiology**

Retinitis pigmentosa is a term used to describe several different inherited disorders of the retina that are characterized by progressive dysfunction involving photoreceptors and often subsequently other cell layers, leading to cell loss and progressive atrophy of several retinal layers. (Ryan, 1994:335) The disease is therefore classified in several different ways. Ideally classification would be dependent on the gene defect present but this is difficult to isolate. Methods of classification depend primarily on the family history and ophthalmologic evaluation.

Genetic inheritance is classified into one of three types, each having many clinical features in common. These types are autosomal dominant (ADRP), autosomal recessive (ARRP) and x-linked recessive (XLRP). The incidence of each type is described in Appendix A. Cases are sometimes sporadic, describing a new mutational event, or simplex, where only one member of a pedigree is affected. Many simplex cases are in fact thought to be due to autosomal recessive inheritance which is difficult to trace. (Ryan,1994:336)

Regardless of the classification the disorder is generally considered to be a dystrophy or genetically determined degeneration. Genetic analysis has shown that the primary defect in most affected individuals is due to a defect in the gene that codes for rhodopsin. This mutation of rhodopsin leads to prolonged dark adaptation problems in subjects and recent studies have shown that “abnormalities in the interactions between rods and cones may underlie the symptoms of poor night vision in some patients”. (Ryan, 1994:336) The disease is described in terms of Type I where the rods are effected earlier than the cones and Type II where the cones are affected earlier than the rods. Atrophy of both the rods and the cones is eventually seen along with migration of pigment epithelial cells into the retina. (Tasman & Jaeger, 1996:153) Histologically studies suggest that pigment deposition in the retina or in the walls of sclerotic retinal vessels come from the

retinal pigment epithelium (RPE). Retinal deterioration is intrinsic to the disease and not necessarily related to pigment deposition, however the contribution pigment makes to loss of retinal function is not known. (Henckenvively et al, 1993: 128)

Patients with RP can also be considered in two large groups. Those where the disease is confined to the eye, such as the subject of this case study, and those where systemic disease is present. These disease types are listed in the section titled "Differential Diagnosis".

Age of onset of the disease varies greatly. This is another method of defining the type of disease present. The disease can present in the following ways:

- **Congenital onset:** as seen in Lebers congenital amaurosis. Onset is at < 1 year of age. Associated signs are nystagmus and poor pupillary reflexes. Usually autosomal recessive inheritance.
- **Childhood onset:** No suspected infant blindness or nystagmus seen. Good visual function in infancy and early childhood.
- **Juvenile onset:** Most common late first or early second decade. All types of inheritance may present in this fashion.
- **Adult onset:** Many types can present as late as the fourth decade, especially autosomal dominant inheritance. Autosomal recessive disease usually presents consistently (age of onset) in siblings.
- **Late onset:** usually autosomal recessive or rarely autosomal dominant disease.

(Ryan, 1994:379)

## **Presenting Clinical Manifestations**

On presentation visual acuity was performed and was 6/6 in both eyes with correction. After examination of the correction being worn it was discovered the patient was highly myopic. This is significant as many forms of RP are associated with myopia. (Ryan, 1994:336) Both eyes were dilated using tropicamide 0.5% and full ocular examination performed.

Dilated fundus examination revealed early posterior subcapsular lens opacities in the right eye, the left lens being clear. Bilateral vitreous opacities were also noted. Posterior vitreous detachment in both eyes was suspected but not confirmed by B-Scan. Both optic discs were tilted but otherwise normal. A vague bulls eye pattern was detected at the macula with the right eye being more defined than the left. No bone spicule pigmentation was seen in either eye.

## **Common Presenting Signs and Symptoms**

One common feature of RP is insidious, progressive loss of visual field. This typically begins with relative scotomata that develop in the midperiphery and enlarge to form a ring scotoma. (Ryan:1994:338) Progressive contraction of visual field leaves a tiny island of central vision which may eventually be lost. (Kanski, 1994:410)

Rarely visual fields will change dramatically over several years or even months. Visual fields usually correspond quite closely with pigmentary changes seen in the fundus. It is worth noting, however, that in Type II disease half of the patients have no intraretinal pigment deposition. (Ryan, 1994:338)

Nyctalopia or night blindness is also a hallmark symptom of RP. This problem typically presents in the first or second decade of life and generally once the visual field has contracted to less than 10 degrees. (Ryan, 1994:336)

Quality of central vision is largely dependent on the type of RP diagnosed. Colour vision generally remains good until vision is disturbed to a level of 6/12 or worse. Unfortunately central and colour vision can be affected by other associated problems including central macular oedema (CMO), diffuse retinal vascular leakage, wrinkling of the internal limiting membrane, macular preretinal fibrosis and RPE defects at the macula or fovea. (Ryan, 1994:341)

Fundus findings in RP are usually bilateral. Kanski describes a classical triad of arteriolar attenuation, retinal bone spicule pigmentation and waxy pallor of optic disc. (1994:410)

Early in the disease common findings are arteriolar narrowing, fine dust-like intraretinal pigmentation, loss of pigment from RPE and a normal optic nerve head. Sometimes macular luster or early premacular fibrosis may be seen. (Ryan, 1994:342)

As the disease progresses “bone spicule” pigmentary changes, due to depigmentation or atrophy of retinal pigment epithelium, irregular pigment clumps, initially in the mid-peripheral retina are seen. These extend anteriorly and posteriorly giving a ring like scotoma in visual field. At this stage the optic nerve becomes pale and waxy. (Kanski, 1994:411)

In advanced states unmasked choroidal blood vessels are observed giving the fundus a tessellated appearance. Prominent arteriolar attenuation is also present. (Kanski, 1994:411 )

Other findings detailed by Ryan (1994:341) and Freeman (1998:1) include:

- *Vitreous*: fine, dust-like pigmented cells or cotton ball like opacities, posterior vitreous detachment (PVD), spindle shaped condensations and asteroid hyalosis in advanced disease.
- *Anterior segment*: posterior subcapsular cataract (prevalence ADRP=52%, ARRP=39%, XLRP=72%), keratoconus, chronic open angle glaucoma.
- *Refractive associations*: high myopia and astigmatism. In patients with high myopia, such as the subject of this case study, often the typical findings of myopia delay the appreciation of other abnormalities in the fundus.
- *Other*: optic nerve drusen, periphery choroidal atrophy

Electrophysiologically the electrooculograph (EOG) and electroretinogram (ERG) are primarily affected in patients with RP. (Pavan-Langston, 1996:172)

EOG measures slow changes in the resting retinal potential and detects disease that interferes with the functional interplay between the RPE and the photoreceptors. ERG reflects the chain of graded electric responses from each layer of the retina. Rod mediated and cone mediated responses can be differentiated by the ERG. This is a useful tool in measuring retinal function loss before ophthalmoscopic changes are evident or vision is lost. (Pavan-Langston, 1996:157). In late stages of the disease the ERG is usually nonrecordable. (Cullom & Chang, 1994:337)

### **Immune Responses**

Retinitis pigmentosa is a genetically inherited dystrophy or degeneration of the retina. Research into the disease through text books, journals and on the internet has not revealed any obvious immune system involvement in the pathophysiology or the treatment of the disease.

## **Differential Diagnosis**

Differential diagnosis for retinitis pigmentosa are diverse in nature. Other retinal degenerations, dystrophies or diseases may be mistaken for RP. According to Cullom & Chang (1994:338) these include:

- Pigmented paravenous retinochoroidal atrophy
- Gyrate atrophy
- Choroideremia
- Resolved exudative retinal detachment
- Congenital stationary night blindness
- Pigment migration into the sensory retina due to blunt trauma or uveitis

Other diagnosis which may be mistaken for RP which are acquired due to systemic disease or treatment with systemic drugs include:

- Phenothiazine toxicity
- Syphilis
- Congenital rubella
- Vitamin A deficiency

RP may also present clinically as part of a number of recognized syndromes.

Some of the more common syndromes associated with RP according to Tasman & Jaeger (1996:154 – 155) :

- Laurence-Moon-Biedl-Bardet Syndrome
- Refsum's Syndrome
- Bassen-Kornzweig Syndrome
- Speilmeyer-Vogt-Batten-Mayou Syndrome
- Progressive External Ophthalmoplegia

## **Diagnostic Methods**

The patient in this case study had a comprehensive medical, family and ocular history taken. Visual acuity and refraction was assessed prior to dilated fundus examination being performed. The subject was referred to a specialist clinic with electrophysiological testing in mind.

A careful and thorough history and examination are vital in accurate diagnosis of RP. Cullom and Chang (1994:344) outline the following methods for a comprehensive assessment of a patient who is suspected of having RP.

1. Medical and ocular history
2. Drug history
3. Family history
4. Ophthalmoscopic examination including visual acuity, refraction, colour vision testing, pupil reflex response, slit lamp examination, intraocular pressure
5. Formal visual field testing
6. Electroretinogram (ERG) and dark adaptation studies
7. Dilated fundus examination, photographs and fluorescein angiography
8. If patient is male and type of inheritance unknown, examination of the mother and ERG
9. If neurological abnormalities such as ataxia, polyneuropathy, deafness or anosmia are present obtain a fasting serum phytanic acid level to rule out Refsum's disease.

In Type I disease careful family history, clinical examination and examination of family members to detect carrier state is the first step in diagnosis. Carrier state is indicated by isolated patches of bone spicules, splotchy pigmentation of the RPE or a bronze sheen in the macula. These findings along with positive ERG findings can confirm diagnosis. Type II disease requires further investigation for positive diagnosis including some or all of the above mentioned examinations. (Tasman & Jaeger, 1996:153)

## **Management**

There is no recognized, clinically tested treatment for RP. Most sources indicate little intervention beyond provision of low vision aids and genetic counselling. (Pavan-Langston, 1996:172) The opportunity to provide patients with understanding of their disease and information on possible future levels of visual function should be taken. Associated refractive errors can be treated to maximise existing vision for the patient, as is the case with the subject of this study. Tinted lenses may also provide comfort for patients, particularly in the out of doors. Particular devices to aid vision in low light areas are available such as the monocular hand-held illuminator or a broad beamed flashlight, these prove useful to RP patients whose night vision is impaired. (Ryan, 1994:455)

Treatment can be administered to alleviate the symptoms of complications of the disease including:

- If significant cataract – extraction and IOL
- Treat CMO with acetazolamide (250mg/day)

(Tasman & Jaeger, 1996:153)

If RP is diagnosed as part of a syndrome there may be treatment available for other systemic malfunctions. In this case study, however the RP was confined to ocular symptoms only.

A search of the internet revealed some exciting research underway into treatment of RP. Much of this work centres around the use of vitamin A and retinal transplantation. Daily administration of 15 000 IU of vitamin A slowed rate of deterioration in clinical trials but no demonstrable benefit for visual acuity or visual fields was seen. (Tasman & Jaeger, 1996:154) A list of other research projects found on the internet can be found in Appendix B.

## **Pharmacology**

An important part of the assessment of the subject of this case study was dilated fundus examination. Tropicamide 0.5% one drop every ten minutes for half an hour was used to dilate the pupil in order to examine the retina. **Tropicamide** is an anticholinergic agent which blocks the response of the sphincter muscle of the iris and the ciliary muscle to cholinergic stimulation, dilating the pupil (mydriasis). The drug acts rapidly and has a short duration of action. It should not be used in cases of known sensitivity or in patients with narrow angle glaucoma. Adverse reactions may include rise in intraocular pressure; transient stinging, dryness of mouth, blurred vision and photophobia with or without corneal staining, tachycardia, headache, parasympathetic stimulation or allergic reaction. Specific reactions in children may include behavioural disturbance, psychotic reactions and cardiorespiratory collapse. . (Badewotz-Dodd, 1997:642,636 & 819)

One of the aims of management of RP is treating the symptoms of complications. One common complication is cystoid macular oedema (CMO). This can be treated with oral acetazolamide. **Acetazolamide** is a carbonic anhydrase inhibitor. It acts to inhibit fluid secretion and promote diuresis in instances of abnormal fluid retention. In the eye this inhibitory action decreases the secretion of aqueous humor and decreases intraocular pressure. The use of this drug is contraindicated in patients with marked liver or kidney dysfunction, suprarenal gland failure, hyperchloraemic acidosis or hypersensitivity to sulphonamides. Adverse reactions during short term therapy are minimal however due to their nature remain significant. These include paraesthesiae, particularly a tingling in the extremities, some anorexia, polyuria, polydipsia, flushing, thirst, headaches, dizziness, fatigue, irritability and occasional instances of drowsiness and confusion. Acetazolamide is given orally in the treatment of CMO, 250mg per day. . (Badewotz-Dodd, 1997:642,636 & 819)

One of the possible treatments of RP currently being researched is the administration of Vitamin A. **Vitamin A**, a fat soluble vitamin, is an essential part of daily vitamin intake. The recommended daily allowance of vitamin A is 2 500 IU per day. Excess vitamin A may cause birth defects if used in pregnancy. Adverse reaction to vitamin A is carotenaemia. (Badewotz-Dodd, 1997:642,636 & 819)

### **Complications**

Complications of RP as discussed previously result from the disease process and include loss of functional vision as well as those treatable conditions secondary to the disease including posterior sub-capsular cataract (PSCC), cystoid macular oedema (CMO) and preretinal membrane.

PSCC can be treated successfully by cataract extraction and lens implant, a commonly performed procedure today. This enables maximization of the existing retinal vision.

Treatment of CMO is currently confined to the prescription of acetazolamide, with varying degrees of success.

Preretinal membrane can be treated successfully by surgical removal in conjunction with par plana vitrectomy. This treatment also aims to improve residual macular function.

## **Legal Considerations/Ethical considerations**

The most important legal consideration is informed consent. This is significant in this case due to the patient being a child, and legally unable to consent for herself. The patient was accompanied by her father who consented to dilation of the eyes for the purpose of examination.

For consent to a procedure to be valid it must consist of three elements:

- a) The consent is freely and voluntarily given;
- b) The consent given is informed; and
- c) The person giving the consent has the legal capacity to do so.

(Staunton & Whyburn, 1997:99 – 102)

“We act on the presumption that the parents will make decisions in the best interests of the child and, on this basis, empower them to do so. However in certain situations, this presumption cannot be sustained.” (Campbell, 1992:73)

Hence the importance of the role of the nurse as an advocate for the child. Value statement number 2 in the Nurses Code of Ethics reads that, “Nurses respect the rights of persons to make informed choices in relation to their care.” (Australian Nursing Council, 1992) This statement refers to the concept of patient advocacy, in ensuring that someone is present to accurately represent the patient’s perspective if they are unable to, such as in the case of a child. Within this concept is the right of the patient to refuse such care if they choose to and the role of the nurse in supporting the patient in that choice. (Australian Nursing Council, 1992)

## **Outcome**

The subject in this case study was referred to a specialist clinic for further investigation. It was likely that electrophysiological testing was to be organized to help confirm or disprove the diagnosis of RP.

Long term outcome for patients diagnosed with RP is largely dependent on the type of disease they have. For patients such as the subject of this case study who have disease which is confined to the eye and not systemic the prognosis is centred on visual function which can effect all activities of daily living.

The amount of visual deterioration is dependent on several factors including inheritance pattern, age of onset and Type I or Type II disease present. Based on the history taken, the patient in this study would be likely to have autosomal dominant inheritance, juvenile onset, Type I disease. Patients with ADRP are more likely to retain good central vision beyond 60 years of age than those with ARRP or XLRP. (Ryan, 1994:341)

About 25% of patients maintain good visual acuity and are able to read throughout their working life, despite unrecordable ERG's and narrowed visual fields. Patients 20 years old and under rarely have visual acuity < 6/60, however by the age of 50 years an appreciable number will be affected to that level. Indirect loss of acuity may occur due to PSCC, CMO or involvement of the fovea. (Kanski, 1994:411)

## **Nursing Care Plan**

The implications of RP to daily living in this case should be examined as short and long term problems. In the short term the impact of the disease on the life of the patient is minimal, as vision is relatively good. As the disease progresses activities of daily living become more greatly affected.

Using the Roper, Logan and Tierney model of nursing, activities of daily living most affected in a patient with RP are the maintenance of a safe environment, communicating, mobilizing and working and playing.

Maintaining a safe environment becomes important as functional vision is lost. Hazards in the home, workplace and community should be identified and methods of preventing accidents explored prior to complete visual loss. Other family members should be educated also about visual impairment and the importance of a controlled and predictable environment, eg: not to move furniture around.

Communicating is an essential part of life and not only includes verbals but non-verbal communication. As visual function is lost, non-verbal communication through facial expression and gesture is limited. The use of touch becomes more important to the visually impaired person.

Mobilising is important in the physical, psychological, economic and social life of the human being. The process of mobilizing changes as vision becomes impaired.

Working and playing is important in providing an income, in creating a purpose in the world, in discovering the satisfaction of achievement and purpose and prevention of boredom.

Each of these activities exist on the dependence/independence continuum. For a child, as in this case study, they are at the stage of emerging independence and this

may be threatened by illness or loss of visual function. Sight can be crucial to independence as an adult. (Roper, Logan & Tierney, 1996:19 –24, 88, 117 – 118)

Nursing care plans for a patient with RP are located in Appendix C.

### **Conclusion**

This case study has examined the history of a patient who presented with suspected retinitis pigmentosa (RP). It has discussed the incidence of RP in the world and in Australia. The pathophysiology of RP in its many forms was explored as well.

The potential and actual presenting symptoms and signs of the subject were outlined along with treatment, both current and future. The pharmacology of medical treatment was also discussed. Expected outcomes and possible complications were discussed including legal and ethical considerations with respect to informed consent and nurse advocacy for the child. Finally nursing care of the patient diagnosed with RP was outlined.

## Reference List

- Australian Nursing Council. 1992, Code of Ethics for Nurses in Australia, Australian Nursing Council, Canberra.
- Badewotz-Dodd, L.H.(Ed) 1997, Mims Annual, Graffin Publishing Co., Ltd., Australia.
- Campbell, A; Gillett, G & Jones, G. 1992, Practical Medical Ethics, Oxford University Press, Auckland.
- Chelva, E; McLaren, T; Kay, s; Collins, D; Black, J & Candy, D. 1992, A Retinitis Pigmentosa Register for Western Australia, Australian and New Zealand Journal of Ophthalmology, Vol 20, No 4, pp 311 - 317
- Cullom, D & Chang, B. 1994, The Wills Eye Manual: Office and Emergency Room Diagnosis and Treatment of Eye Disease, J.B.Lippincott Company, Philadelphia.
- Dee's Retinal Degeneration Research Links,  
<http://www.Netserv.net.au/doonbank/research.html>
- Freeman, W.R. 1998, Practical Atlas of Retinal Disease and Therapy(2<sup>nd</sup> Edition), Lippincott-Raven Publishers, Philadelphia.
- Heckenlively, J R; Abrams, G W; Chuang, E L; Grand, G M G; Green, W R & Guzak, S J. 1993, Basic and Clinical Science Course 1993 – 1994 American Academy of Ophthalmology, Section 12 “Retina and Vitreous”, American Academy of Ophthalmology, San Francisco.
- Kanski, J. 1994, Clinical Ophthalmology, A Systemic Approach (3<sup>rd</sup> Edition), Butterworth-Heineman Ltd, Oxford.
- Pavan-Langston, D. 1996, Manual of Ocular Diagnosis and Therapy, Little, BrownAnd Company (Inc), Boston.
- Roper, N; Logan, W& Tierney, A. 1996, The Elements of Nursing, A Model For Nursing Based on a Model of Living, Churchill Livingstone, New York
- Ryan, S J. 1994, Retina Volume One, Basic Science and Inherited Retina Disease (2<sup>nd</sup> Edition), Mosby, St Louis.
- Staunton, P & Whyburn, B. 1997, Nursing and the Law (4<sup>th</sup> Edition), Harcourt and Brace & Company, Australia.

## Bibliography

- Australian Nursing Council. 1992, Code of Ethics for Nurses in Australia, Australian Nursing Council, Canberra.
- Badewotz-Dodd, L.H.(Ed) 1997, Mims Annual, Graffin Publishing Co., Ltd., Australia.
- Campbell, A; Gillett, G & Jones, G. 1992, Practical Medical Ethics, Oxford University Press, Auckland.
- Chelva, E; McLaren, T; Kay, s; Collins, D; Black, J & Candy, D. 1992, A Retinitis Pigmentosa Register for Western Australia, Australian and New Zealand Journal of Ophthalmology, Vol 20, No 4, pp 311 - 317
- Cullom, D & Chang, B. 1994, The Wills Eye Manual: Office and Emergency Room Diagnosis and Treatment of Eye Disease, J.B.Lippincott Company, Philadelphia.
- Dee's Retinal Degeneration Research Links,  
<http://www.Netserv.net.au/doonbank/research.html>
- Freeman, W.R. 1998, Practical Atlas of Retinal Disease and Therapy(2<sup>nd</sup> Edition), Lippincott-Raven Publishers, Philadelphia.
- Hamilton, P.A; Gregson,R & Fish, G.A. (Ed) 1998, Text Atlas of the Retina, Martin Dunitz, London.
- Heckenlively, J R; Abrams, G W; Chuang, E L; Grand, G M G; Green, W R & Guzak, S J. 1993, Basic and Clinical Science Course 1993 – 1994 American Academy of Ophthalmology, Section 12 “Retina and Vitreous”, American Academy of Ophthalmology, San Francisco.
- Kanski, J. 1994, Clinical Ophthalmology, A Systemic Approach (3<sup>rd</sup> Edition), Butterworth-Heinemann Ltd, Oxford.
- Najera, C. 1995, Epidemiology of retinitis pigmentosa in the Valencian community (Spain), Genetic Epidemiology, Vol 12, No 1, pp 37 – 46.
- Pavan-Langston, D. 1996, Manual of Ocular Diagnosis and Therapy, Little, BrownAnd Company (Inc), Boston.

Roper, N; Logan, W& Tierney, A. 1996, The Elements of Nursing, A Model For Nursing Based on a Model of Living, Churchill Livingstone, New York

Ryan, S J. 1994, Retina Volume One, Basic Science and Inherited Retina Disease (2<sup>nd</sup> Edition), Mosby, St Louis.

Staunton, P & Whyburn, B. 1997, Nursing and the Law (4<sup>th</sup> Edition), Harcourt and Brace & Company, Australia.

## Appendix A

<b>Type of Inheritance</b>	<b>Approximate Incidence of Occurrence</b>
Autosomal Dominant RP	16% of cases
Autosomal Recessive RP	41% of cases
X-Linked Recessive RP	6 – 9% of cases

<b>Type Group</b>	<b>Electrophysiological Presentation</b>
Type I	Rods affected > cones
Type II	Cones affected > rods

## **Appendix B**

### **Australia**

- NSW Retinal Dystrophy Research Centre – Department of Anatomy and Histology and the Institute for Biomedical Research, University of Sydney

### **Germany**

- Sub Retinal Implant Project, University Eye Hospital, Tübingen
- Retina Implant, University of Bonn, Department of Computer Science

### **Japan**

- Hybrid Retinal Implant (Implantable Artificial Retina), Nagoya University

### **New Zealand**

- The Molecular Genetics of Retinitis Pigmentosa, University of Otago, Dunedin, New Zealand

### **South Africa**

- Molecular Genetics of Familial Retinal Degenerative Disorder In Southern Africa, Department of Human Genetics at the University of Cape Town

### **United States of America**

- Artificial Vision Research, Dr Mark Humayun, Dr Wentai Lui and Elliot McGucken
- Centre for Retinal Research, Columbia-Presbyterian Medical Centre
- Centre for the Study of Macular Degeneration, University of California, Santa Barbara
- The Retinal Implant Project, Massachusetts Eye and Ear Infirmary and the Massachusetts Institute of Technology
- The Retinal Prosthesis Project, University of North Carolina
- Retinal Transplants, University of Louisville
- Centre for Inherited Retinal and Macular Degenerative Disease, University of Michigan, W K Kellogg Centre
- Human Genetics Centre and Ophthalmology and Visual Science, University of Texas Health Science Centre.

(Dee's Retinal Degeneration Research Links,  
<http://www.Netserv.net.au/doonbank/research.html>)