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Research Article

# Clinical Diagnostic Dilemma between Reis Buckler and Thiel Behnke Corneal Dystrophy

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## INTRODUCTION

Thiel Behnke corneal dystrophy (Corneal dystrophy of Bowman's layer type II / honeycomb dystrophy)[1] is frequently confused with the hereditary epithelial stromal dystrophy Reis Bucklers. Although histopathological study and characteristic curly fibres on electron microscopy can lead to an unequivocal confirmation , there remains a clinical diagnostic dilemma between these two epithelial stromal dystrophy due to the similarities in symptoms , signs and genetic mutation. In this report , we present a case of biopsy proven Thiel Behnke dystrophy with recurrence in graft of right eye . In this case, we have tried to highlight the importance of extensive clinical examination including a proper clinical history along with the additional benefit of ASOCT and confocal microscopy as diagnostic tool to identify the subtle changes in the corneal anatomy which can help to reduce the diagnostic dilemma.

## CASE PRESENTATION

30 year old male presented with gradual progressive diminution of vision in both eyes since 6 years of age with photophobia and few episodes of watering and pain.

No history of ocular trauma , floaters or coloured halos were elicited.

There was no history of any significant systemic morbidities.

He had undergone both eye phototherapeutic keratectomy (PTK) 3 years ago with pre operative and postoperative visual acuity of 20/30 in both eyes and had undergone superficial anterior lamellar keratoplasty(SALK) in right eye 2 years ago with a pre operative vision of 20/30 and post operative visual acuity of 20/20.

Family pedigree chart (Figure 1) revealed an autosomal dominant inheritance of the disease.

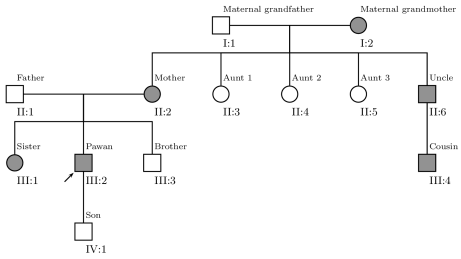
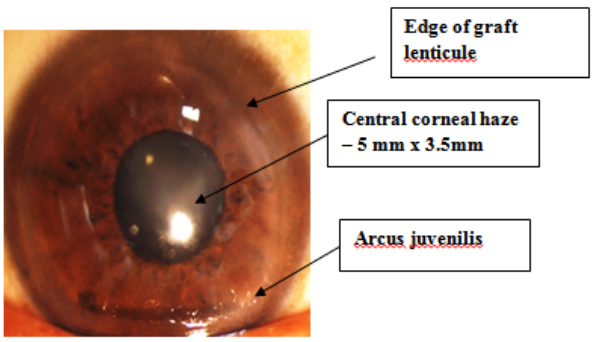



**FIGURE 1 :** Family pedigree chart pointing towards an autosomal dominant inheritance.

Ocular examination revealed best corrected visual acuity (BCVA) of 20/30 in both eye with an IOP of 14 mm Hg .

Slit lamp examination revealed a graft of 9 mm diameter with clarity of grade 4 [2] and a clear interface with a central superficial haze of 5 mm x 3.5 mm and a temporal circumferential haze in right eye (Figure 2 ). Left eye demonstrated diffuse multiple symmetrical honeycomb opacities of 10 mm x 9 mm with clear area between the haze and the limbus and forming a saw tooth pattern on a slit (Figure 3 ).




**Slit lamp examination of RE**

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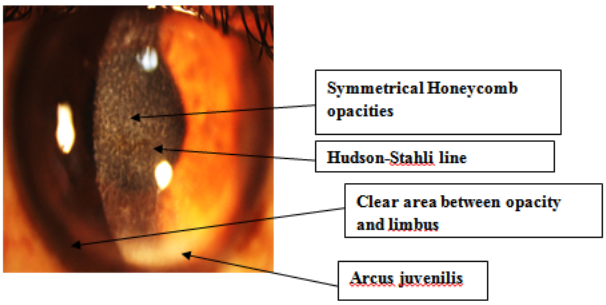
Figure 2

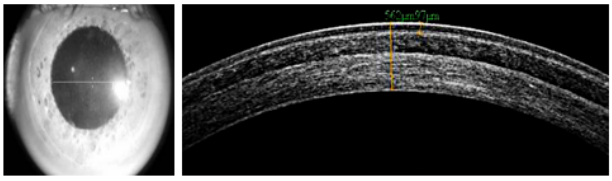



Slit lamp examination of LE

Figure 3

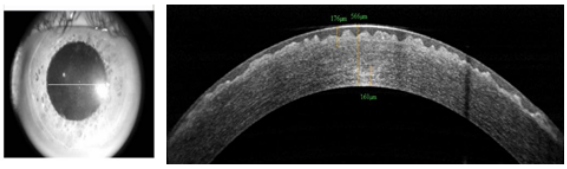
Rest of the anterior segment as well as posterior segment were within normal limits.

ASOCT of right eye demonstrated a CCT of 562µm with depth of recurrence of 97 µm (Figure 4) and LE had a CCT of 566 µm and depth of anterior involvement of 176 µm with saw-tooth pattern of dense hyper-reflective material at bowman's layer with mild posterior stromal haze (Figure 5).



ASOCT OF RE -Anterior graft well attached with no interface haze with mild posterior stromal haze with recurrence in the graft .

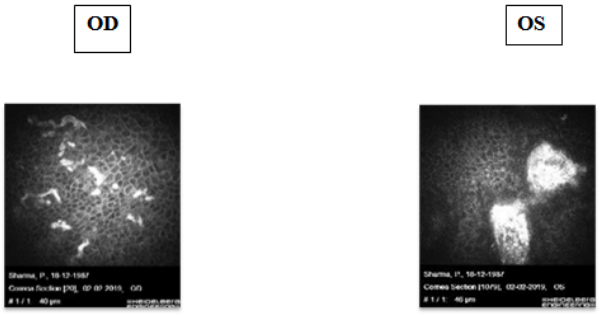
Figure 4



ASOCT OF LE - Saw-tooth pattern of dense hyper-reflective material at bowman's layer with mild posterior stromal haze

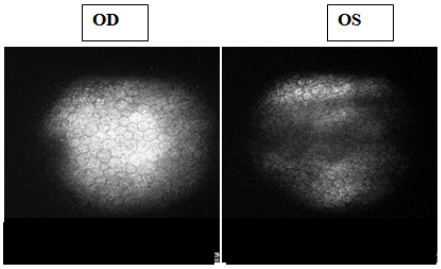
Figure 5

Confocal microscopy revealed wave like deposition of hyper-reflective material replacing the Bowman's membrane at epithelial-Bowman membrane junction in left eye with a normal endothelium in both eyes (Figure 6 and 7)




Confocal microscopy - Abnormal hyperreflective material with homogeneous reflectivity, round edges, and dark shadows noted in epithelial- Bowman's layer junction

Figure 6




Confocal microscopy – BE normal endothelium

Figure 7

Based on clinical examination and investigation findings , we formulated a diagnosis of epithelial stromal dystrophy (? Thiel Behnke corneal dystrophy) with BE operated PTK with RE operated SALK with recurrence in the graft.

# DISCUSSION

The hereditary Thiel-Behnke corneal dystrophy, first described in 1967[3], is frequently confused with Reis-Bucklers dystrophy. Both of them belong to the epithelial stromal TGFß1 dystrophy according to International committee for classification of corneal dystrophy( IC3D classification) [4] and have autosomal dominant inheritance. In our case, we have tried to evaluate all the clinical and investigative findings extensively as per IC3D description and reached our diagnosis.

Table 1: depicts the symptoms of Reis Buckler dystrophy and Thiel Behnke dystrophy and its presence / absence in our case .

| SYMPTOMS | RBCD | TBCD |
|---|---|---|
| Age of onset | 1st and 2nd decade (√) | 1st and 2nd decade (√) |
| Visual acuity | Early and marked visual loss (X) | Reduced later in life (√) |
| Recurrent episodes of pain | Present (√) | Present (√) |
| Family history | AD(√) | AD(√) |

| | | |
|---|---|---|
| **Limbus involvement** | Present in later stages (X) | 1-2 mm clear margin adjacent to limbus (√) |

Figure 8: compares the slit lamp images of Reis Buckler dystrophy and Thiel Behnke dystrophy as per IC3D 2015 with the images in our case.

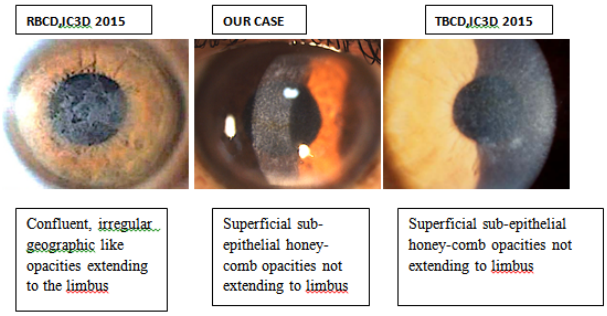



Figure 9: compares the characteristic ASOCT images of Reis Buckler dystrophy and Thiel Behnke dystrophy with the images in our case.

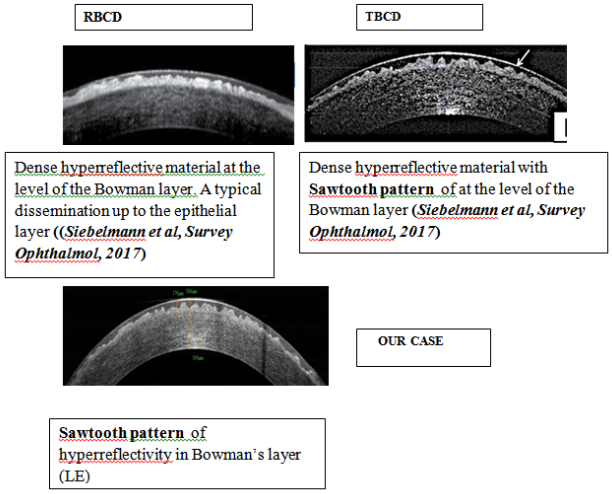
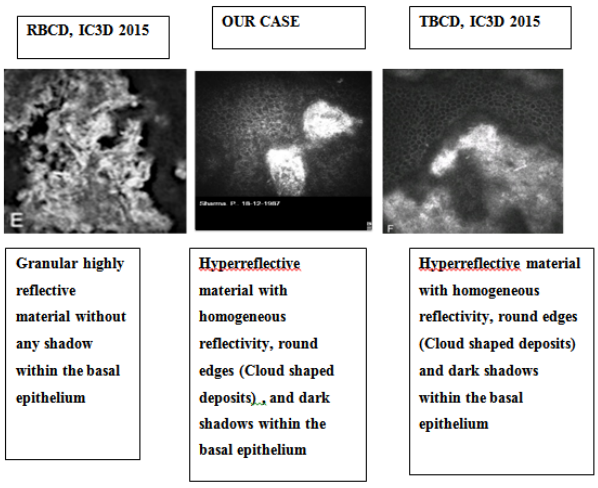



Figure 10 compares the characteristic confocal microscopy images of basal epithelium of Reis Buckler dystrophy and Thiel Behnke dystrophy respectively with the images in our case.




Although genetic, electron microscopy and histopathological examination s lead to a confirmatory diagnosis , an extensive clinical examination with AS-OCT and confocal microscopic examination can point towards an accurate diagnosis.

Left eye Superficial Anterior Lamellar Keratoplasty (SALK) was performed and the histopathological diagnosis corroborated with our clinical diagnosis.

## REFERENCES

1. Küchle M, Green WR, Völcker HE, Barraquer J. Reevaluation of corneal dystrophies of Bowman's layer and the anterior stroma (Reis-Bücklers and Thiel-Behnke types): a light and electron microscopic study of eight corneas and a review of the literature. Cornea. 1995;14(4):333-354.
2. Vanathi M, Sharma N, Sinha R, Tandon R, Titiyal JS, Vajpayee RB. Indications and outcome of repeat penetrating keratoplasty in India. BMC Ophthalmol. 2005;5:26.
3. Thiel HJ, Behnke H. A hitherto unknown subepithelial hereditary corneal dystrophy. Klin Monbl Augenheilkd. 1967;150(6):862-874.
4. APA Weiss, Jayne S. MD; Møller, Hans Ulrik MD, PhD; Aldave, Anthony J. MD; Seitz, Berthold MD; Bredrup, Cecilie MD, PhD; Kivelä, Tero MD, FEBO; Munier, Francis L. MD; Rapuano, Christopher J. MD; Nischal, Kanwal K. MD, FRCOphth; Kim, Eung Kweon MD, PhD; Sutphin, John MD; Busin, Massimo MD; Labbé, Antoine MD; Kenyon, Kenneth R. MD; Kinoshita, Shigeru MD, PhD; et al, Lisch, Walter MD IC3D Classification of Corneal Dystrophies—Edition 2, Cornea: February 2015