



# ENDOCRINE DISORDERS IN CHILDREN WITH THALASSEMIA

## MAJOR - A RETROSPECTIVE STUDY

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### INTRODUCTION

- **Beta Thalassemia** represents a group of *recessively inherited* hemoglobin disorders first described by *Cooley and Lee*, where beta globin chain synthesis is decreased resulting in an excess of alpha chains, resulting in severe anemia.
- The estimated prevalence of beta thalassemia is **3-8%** in *India, Pakistan, Bangladesh and China*.
- The combination of regular *blood transfusions* and *chelation therapy* has dramatically increased the life expectancy of thalasseemics into 4th & 5th decades of life.
- But it has led to *iron overload* and *chelation toxicity*, with many complications including – *growth failure, gonadal dysfunction, hypothyroidism, DM etc.*

### AIMS AND OBJECTIVES

- To study the **Incidence and Prevalence** of endocrine disturbances in Thalassemia Major children, receiving regular blood transfusions.
- To study the correlation between-
  - *Age of starting transfusion*
  - *Average Sr. Ferritin levels*
  - *Age of starting chelation***AND** incidence of *Endocrine problems* in these children.

### METHODOLOGY

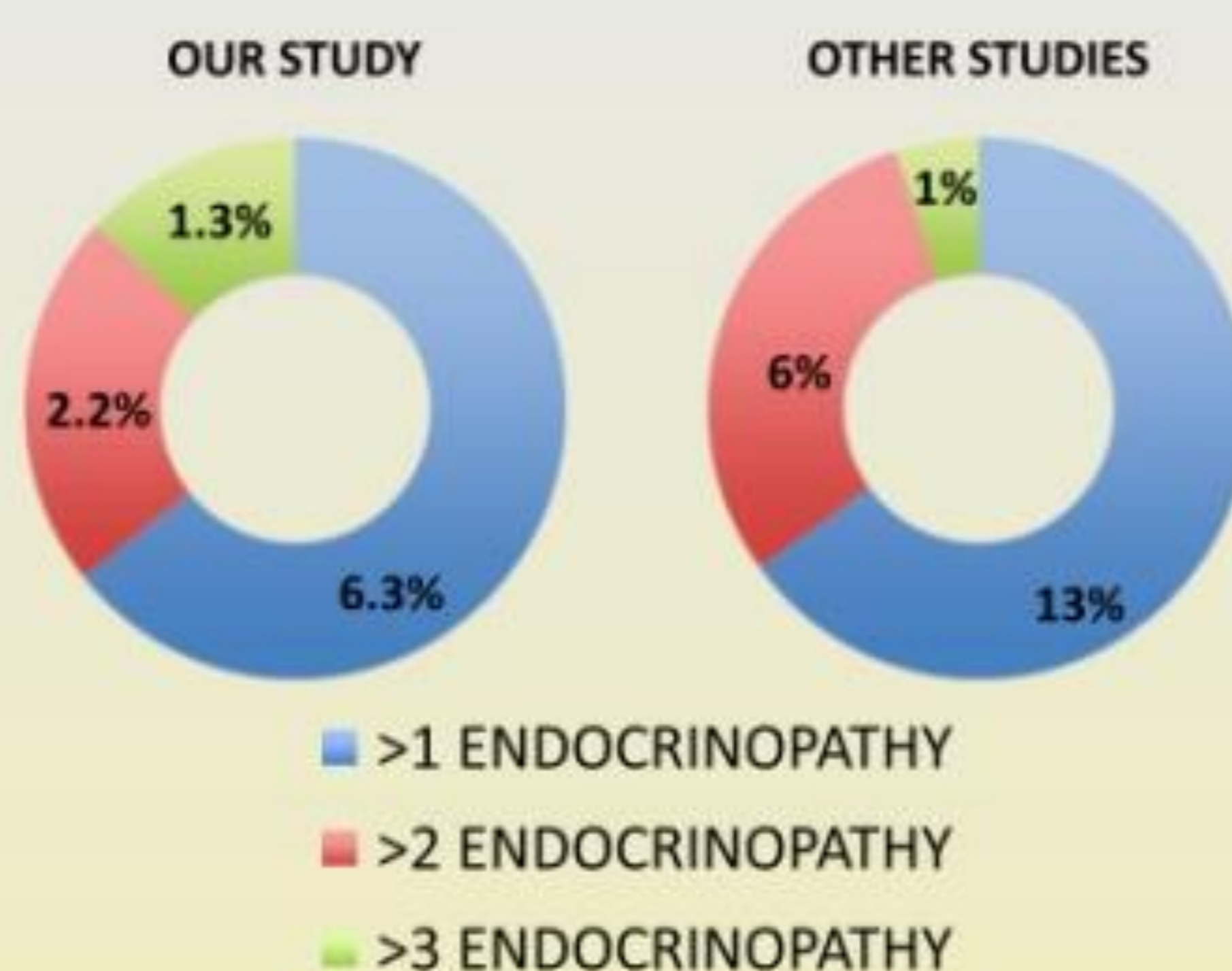
- A *retrospective study* of thalassemia major patients.
- Basic information for each child was determined -
  - Age of presentation, onset of transfusion and chelation.
  - Anthropometric data.
  - Pubertal status- Tanner staging.

### Blood investigations

- Sr. Ferritin and average ferritin levels
- Hemoglobin level
- Sr. Calcium/Phosphorus/Alkaline Phosphatase
- PTH (Parathyroid Hormone)
- Thyroid profile – FT3/FT4/TSH
- OGTT (Glucose tolerance test)
- LH/FSH
- Sr. Estradiol (girls)/Sr. Testosterone (boys)

### RESULTS

- **225** children were included in the study.
- **32.4%** (73/225) children had endocrine complications, while **9.4%** of them had more than one endocrine complication.



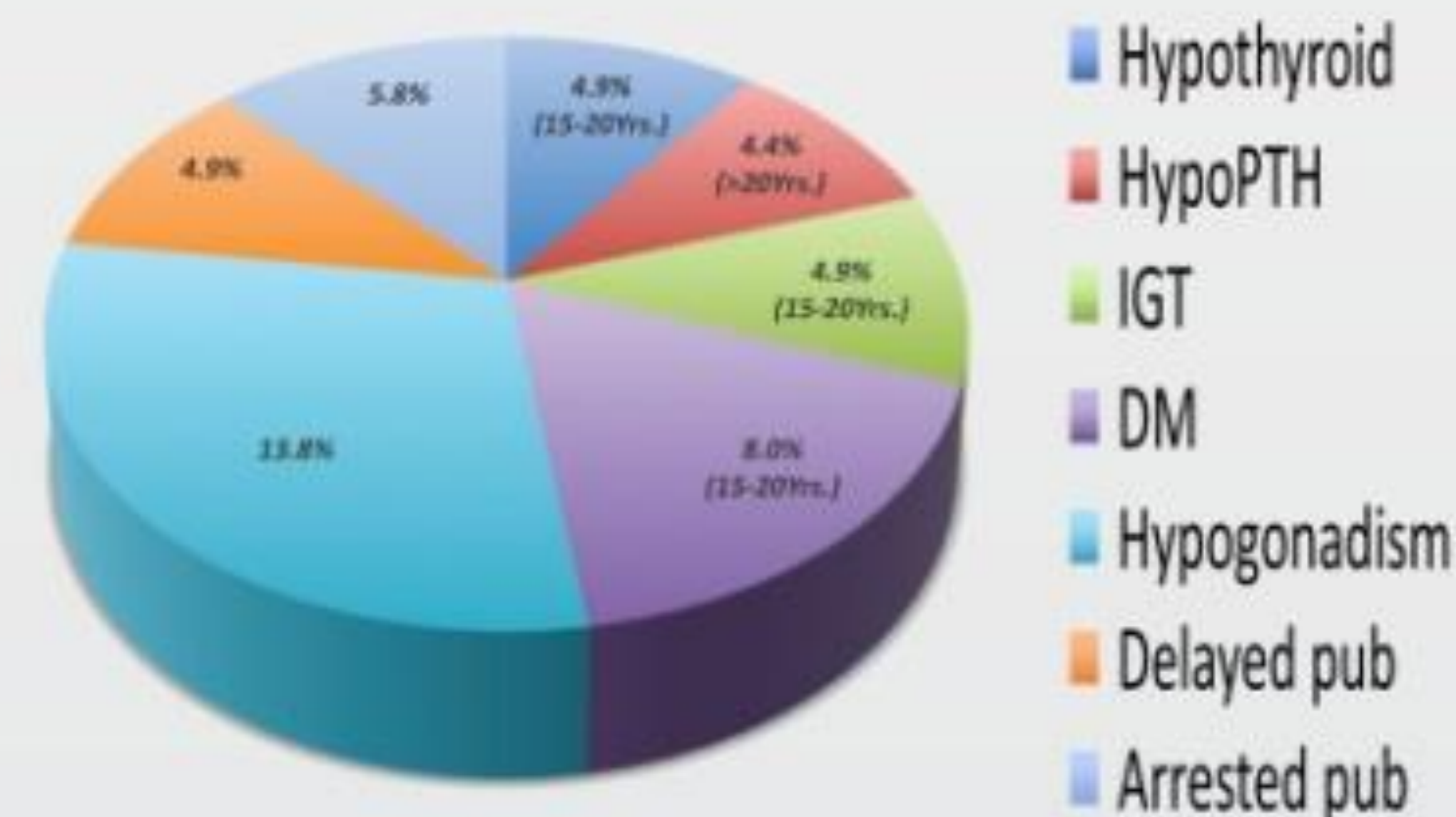
- Endocrine complications increased with **Age** – maximum seen in age group **>15 years**.



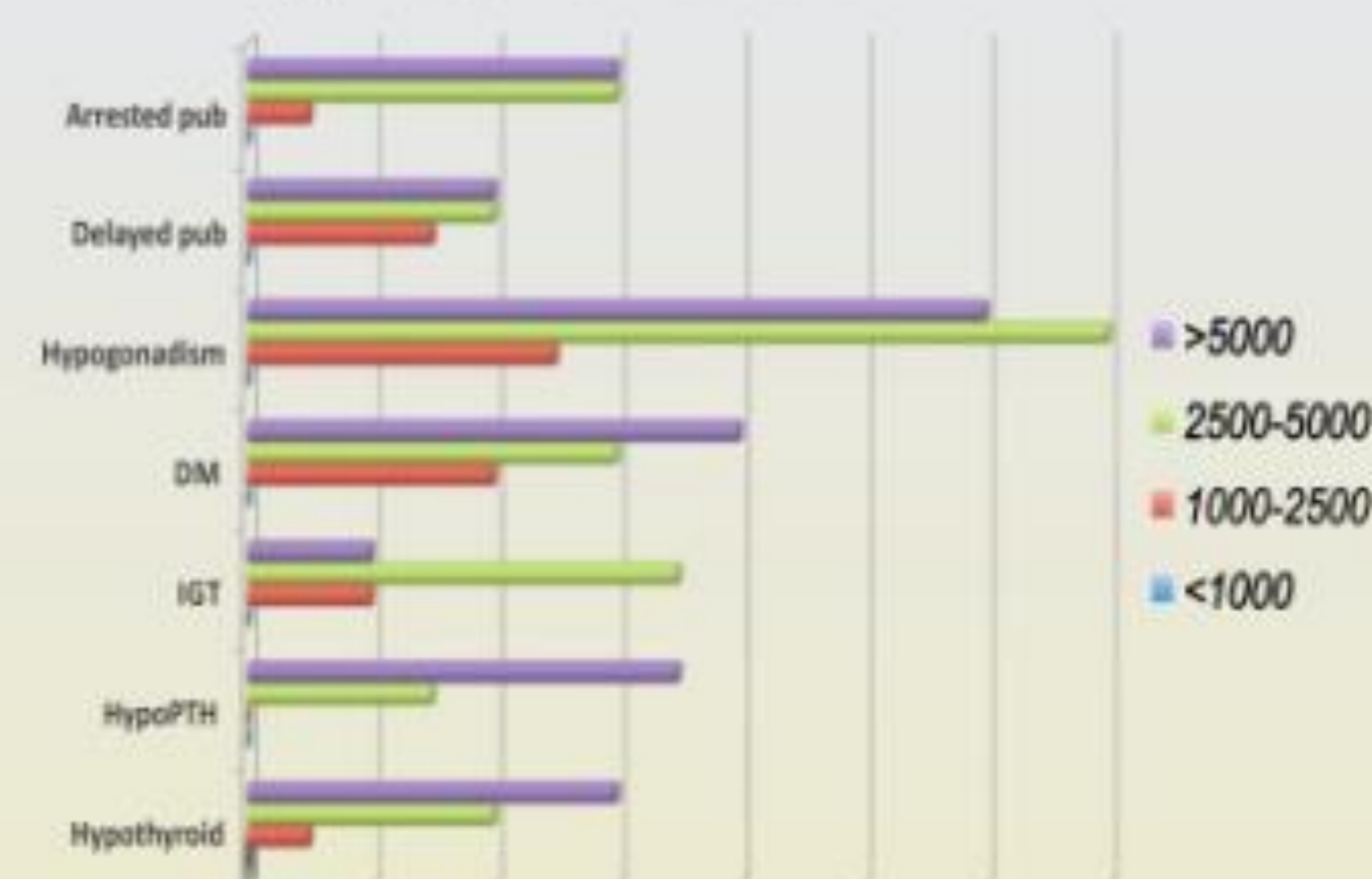
Age and Endocrine Complications

- **No association** was found between *age of onset of transfusion and chelation* with *incidence of endocrine problems*.

### Incidence of different endocrine problems



### Average Ferritin levels and Incidence of Endocrine Problems



	Our study	Inati et al	C K Ong et al	Italian Working Group & Shalitin et al
Hypogonadism	+	-	+	+
Delayed puberty	-	-	-	+
IGT/DM	-	-	-	-
Hypothyroidism	+	-	-	-
HypoPTH	+	-	-	-

### CONCLUSION

Endocrine complications (% in both sexes)	Country					
	Cyprus (435)	Greece (262)	Italy (1861)	Tehran (220)	N.Am (262)	Our study(225)
Hypothyroidism	5.9	4	6.2	7.7	4	4.9
HypoPTH	1.2	4	3.6	7.6	4	4.4
DM/IGT	9.4	5/27	4.9	8.7	5	8/4.9
Hypogonadism/Delayed Pub/Arrested Pub	32.5	42	49	17.5/3 5.1	42	13.8/4.9/5.8

Large scale studies are required to determine **true incidence** of Endocrine complications in Thalassemia.

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• **Conflict of interest: None**

