**Supplementary Figure S1.** Venn diagrams summarizing the overlap of DEGs between OI versus controls and IFAP/KFSD versus controls.



**Supplementary Figure S2.** Principal Component Analysis biplot of PC1 and PC2 is depicted to visualize the degree of similarities among the gene expression profiles of control and patient fibroblasts at basal conditions (nt) and 24 hours after thapsigargin (TG) treatment to induce ER stress in the top panel. The proportion of variance explained by each PC is shown in the screeplot in the bottom panel.



Supplementary Figure S3. The correlation of each sample to all other samples was analysed by Pearson's correlation coefficient.

OI P3_TG -									J		,								_	- 0.962
OI P2_TG -																				
OI P1_TG -																				- 0.95
OI P3_nt -																				- 0.938
OI P2_nt -																				0.950
OI P1_nt -																				- 0.927
KFSD_TG -																				
IFAP_TG -																				- 0.915
KFSD_nt -																				
IFAP_nt -																				- 0.903
control 3_TG -																				
control 2_TG -																				- 0.892
control 1_TG -																				0.99
control 4_nt -																		l		- 0.88
control 3_nt -																				- 0.868
control 2_nt -																				
control 1_nt -																				- 0.856
	11_nt -	l 2_nt -	l 3_nt -	4_nt -	1_TG	2_TG -	3_TG -	IFAP_nt -	KFSD_nt -	IFAP_TG -	KFSD_TG -	OI P1_nt -	OI P2_nt -	OI P3_nt -	OI P1_TG -	OI P2_TG -	OI P3_TG -			
	control 1_nt	control 2_nt	control 3_nt	control 4_nt	control 1_TG	control 2_TG	control 3_TG	IF/	KFS	IFA	KFSI	O	O F	IO 1	OI P	OI P	OI P			

all present genes (15031)



l -3 -2 -1 0 1 2 3 log2 difference

Supplementary Figure S4. Heatmap depicting expression levels of ATF6-dependent genes at basal conditions.



	EMSY STAG1 ERO1B
Image: second	TNKS2
Image: second	SEL1L TRPC1 LMBRD1
Image: state	ERO1A WIPI1
Image: state	OSBP COPB1
Image: second	SEC62 SLC30A5
Image: second	CANX SEC31A
	TBC1D23 HYOU1
	ZFHX3 SLC35E1
	DNAJC10 DHX40
	BRD2 SEC61A1
	SYVN1 SEC24D LMAN1
	SEC23A GBF1
	RFTN2
	DERL2 SRP19 DNAJB11
	GPBP1 DDIT3 HSP90B1
	PDIA3
	HSPA5 CALR
	PLPP5 NANS TMEM263
	ARF4 LRRC59
	SRPRB MSTO1
	TOP1 RPN1
	STX5 JOSD1
	SEC23B STT3A
	XBP1 TMEM125
	SRPRA DDOST TRIM39
	GCC1 OS9
	HERPUD1 PDIA4
	CRELD2 ICA1

Color Key



Supplementary Figure S5. Heatmap depicting expression levels of OASIS-dependent genes at basal conditions.



## Color Key



## Supplementary Figure S6. Heatmap depicting expression levels of BBF2H7-dependent genes at basal conditions.



Color Key



**Supplementary Figure S7:** Heatmap depicting expression levels of genes that are transcriptionally regulated by SREBP at basal conditions (nt: no treatment) and in the presence of thapsigargin (TG).





Supplementary Figure S8. A simplified diagram of cholesterol biosynthesis is depicted. The cellular content of sterol metabolites was determined by GC-MS/MS and expressed as ratios. Four independent measurements were performed per subject, and t-tests were performed for statistical analysis. Data are expressed as mean ± SEM.



1000

500·

0

control

οι

IFAP/KFSD













ratio



## 7-dehydrodesmosterol : lanosterol