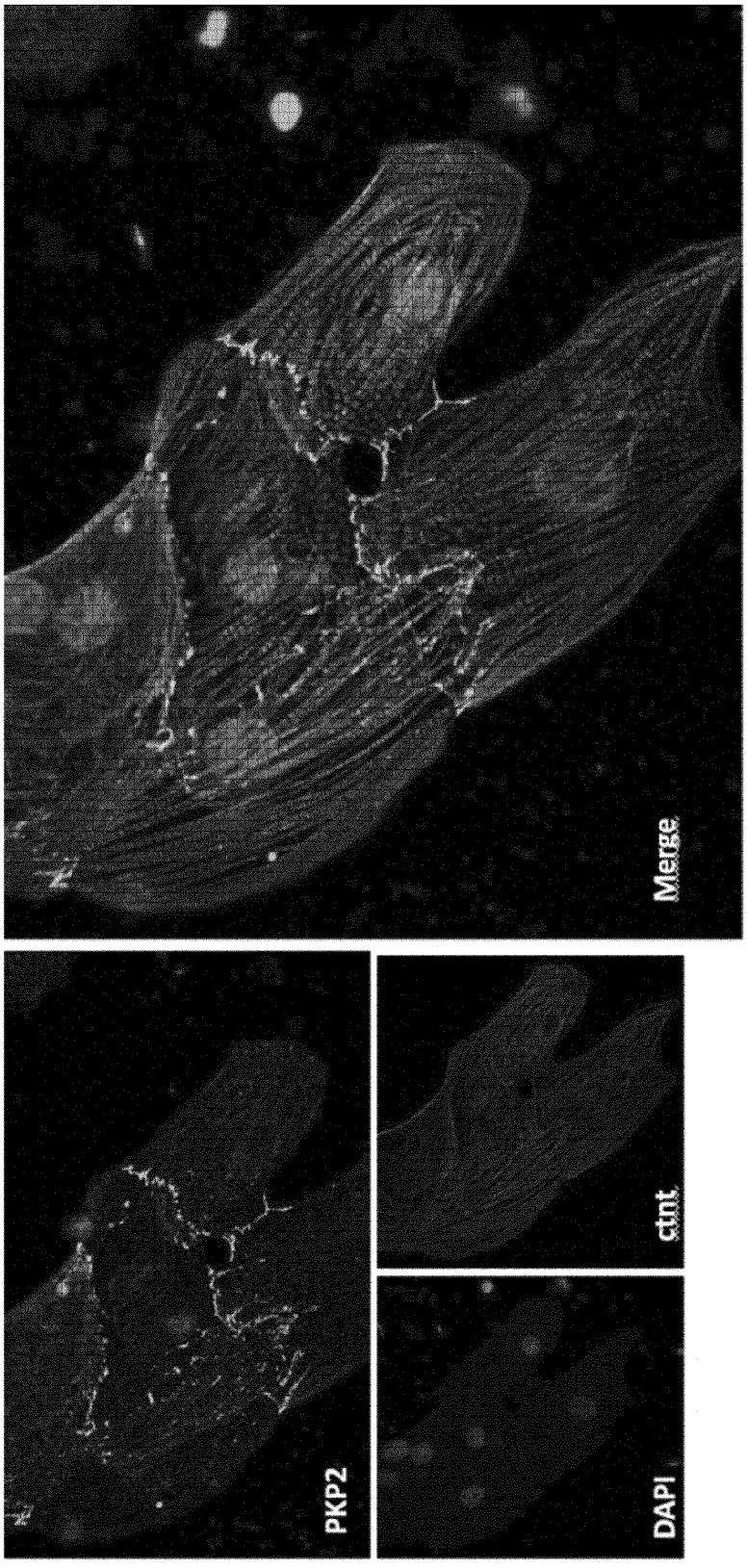

FIG. 1

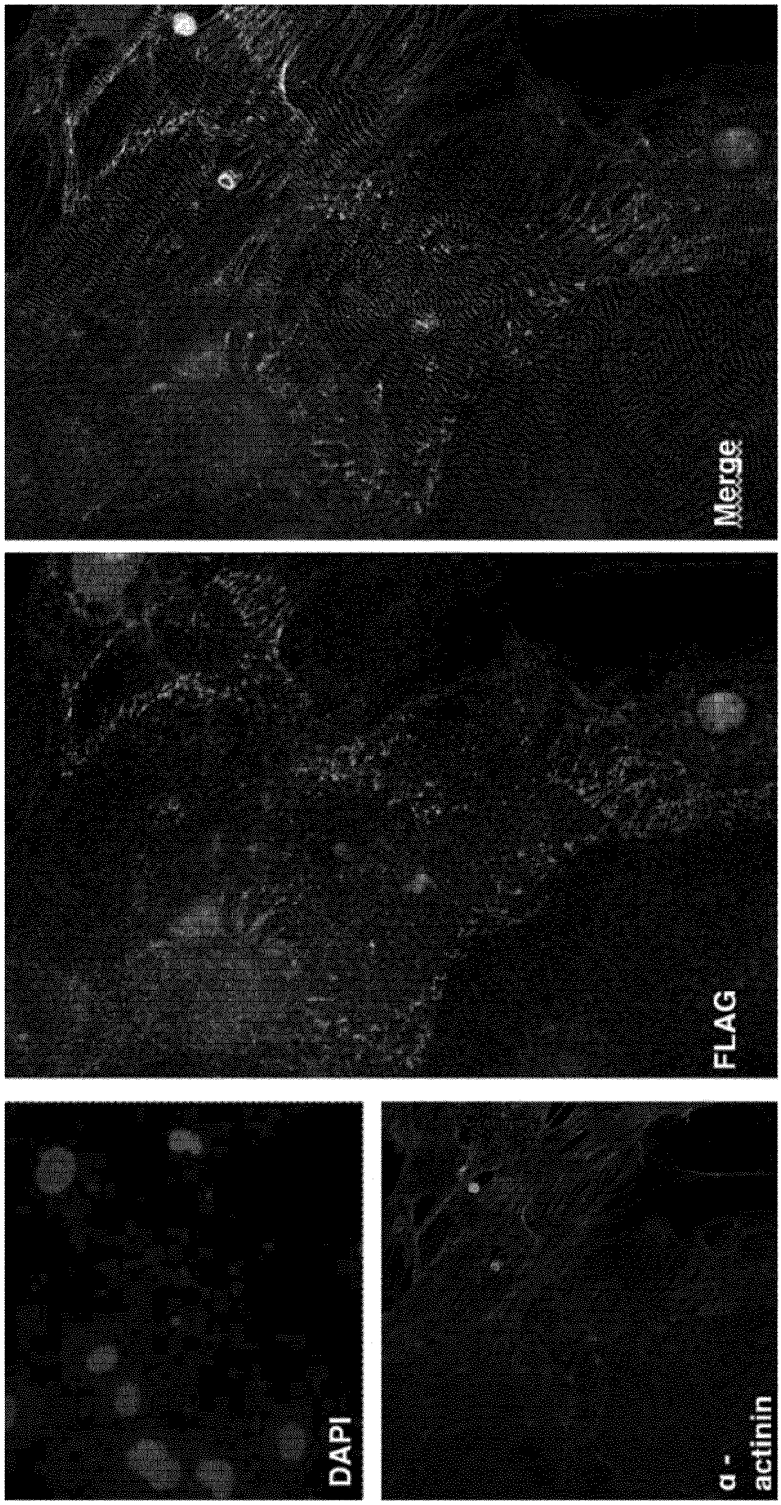

FIG. 2

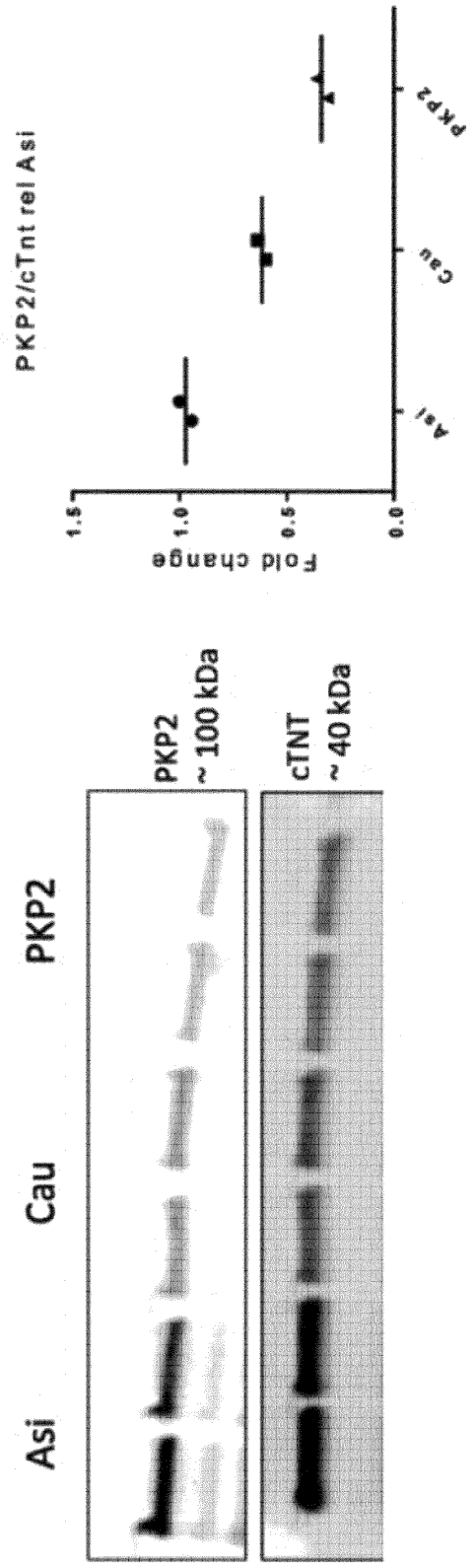
PKP2/cTnt rel Asi
Fold change
1.5
1.0
0.5
0.0
Asi
Cau
PKP2
Asi
Cau
PKP2
~ 100 kDa
cTNT
~ 40 kDa

FIG. 3

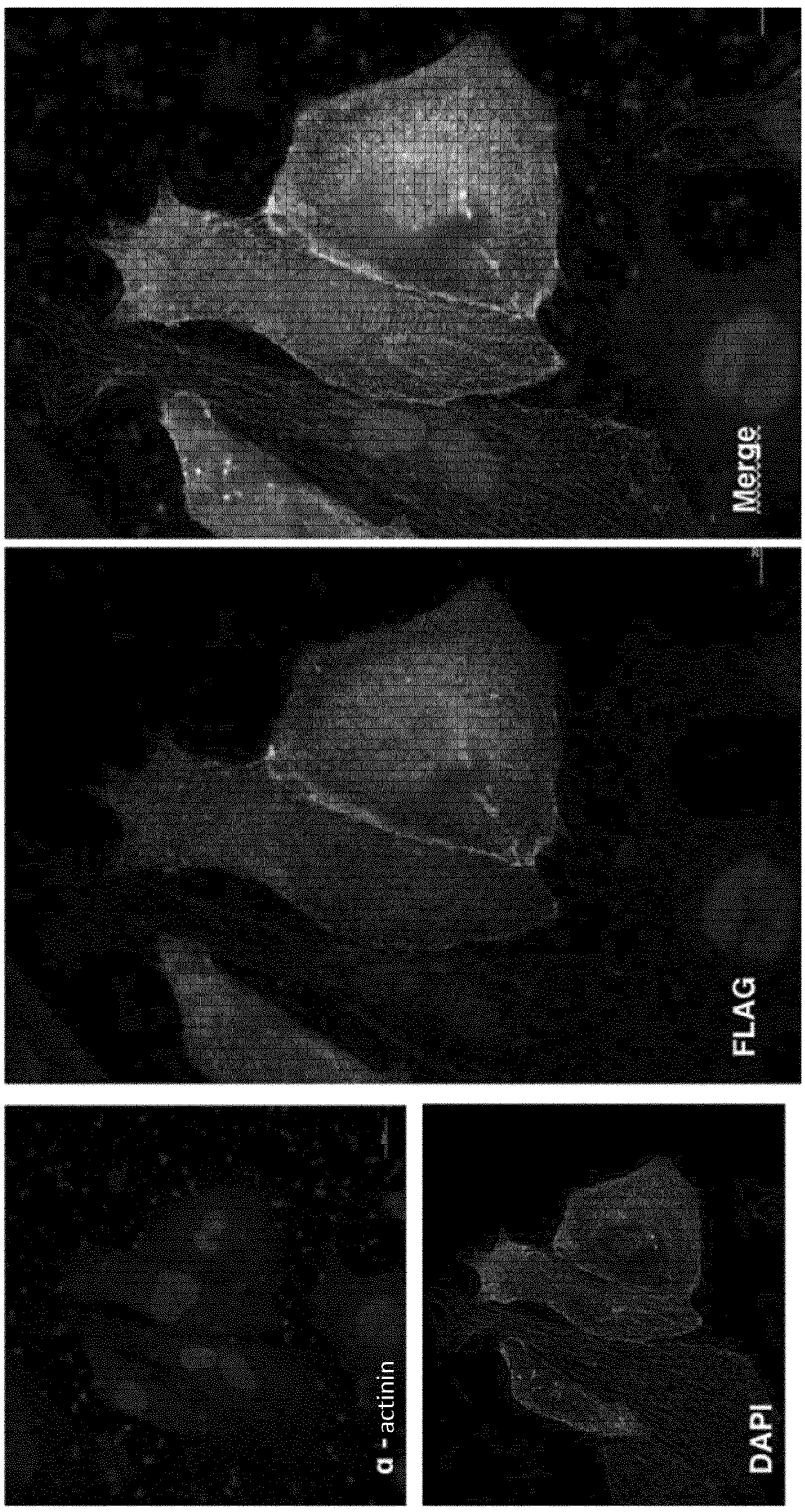

FIG. 4

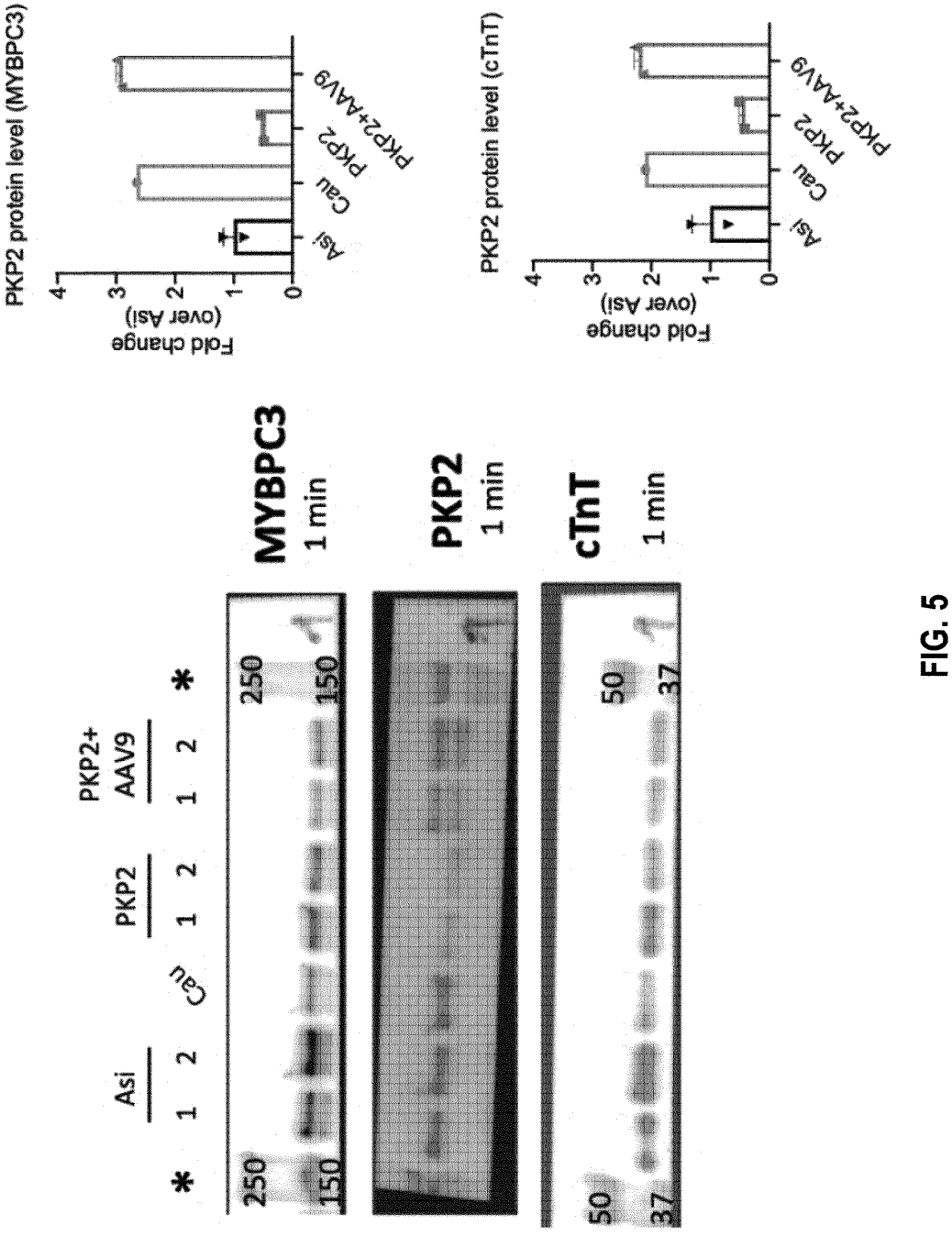
PKP2 protein level (MYBPC3)
Fold change (over Asi)
Asi
Cau
PKP2
PKP2+AAV9
PKP2 protein level (cTnT)
Fold change (over Asi)
Asi
Cau
PKP2
PKP2+AAV9
MYBPC3
1 min
PKP2
1 min
cTnT
1 min
Asi
Cau
PKP2
PKP2+ AAV9
250
150
50
37

FIG. 5

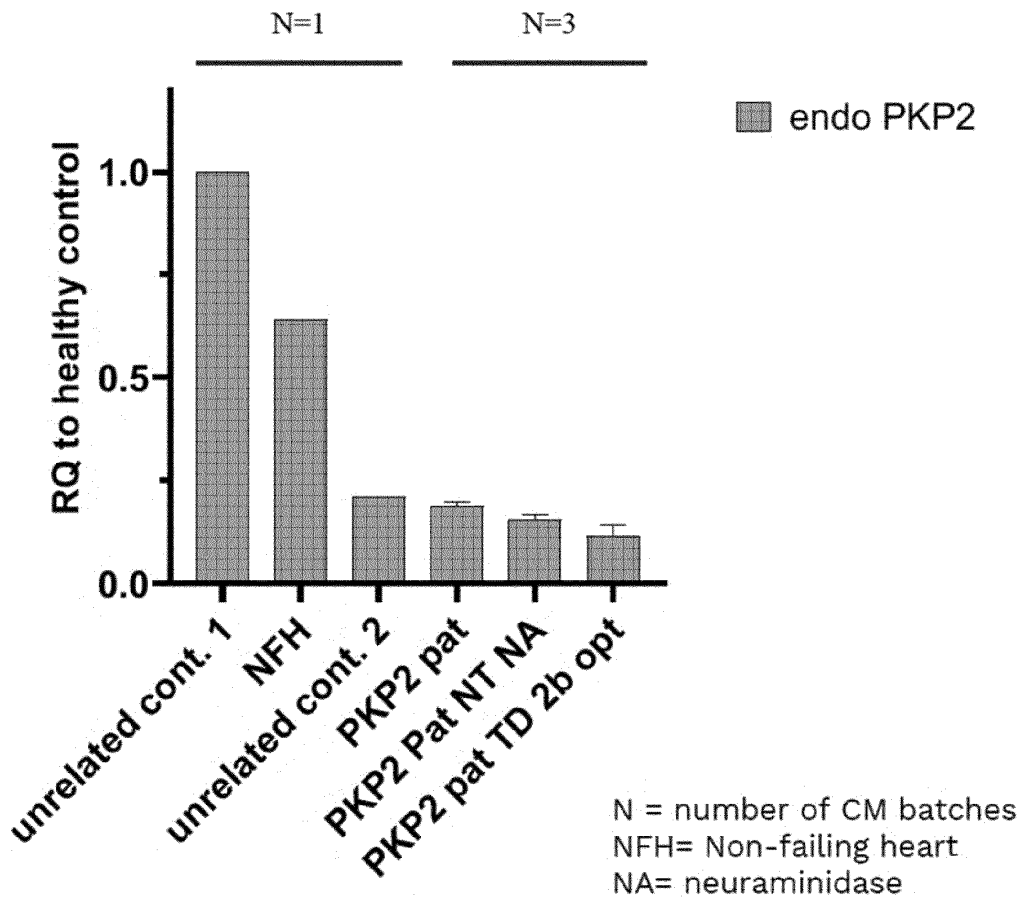
N=1
N=3
endo PKP2
RQ to healthy control
1.0
0.5
0.0
unrelated cont. 1
NFH
unrelated cont. 2
PKP2 pat
PKP2 Pat NT NA
PKP2 pat TD 2b opt
N = number of CM batches
NFH= Non-failing heart
NA= neuraminidase

FIG. 6

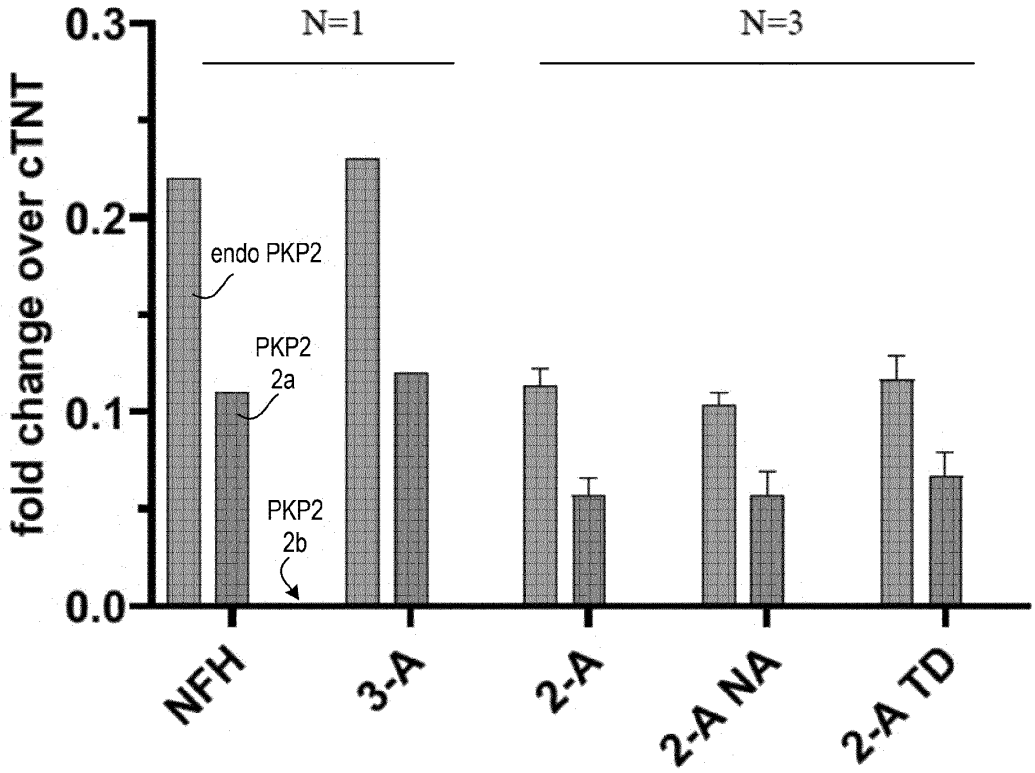
0.3
0.2
0.1
0.0
fold change over cTNT
N=1
N=3
endo PKP2
PKP2
2a
PKP2
2b
NFH
3-A
2-A
2-A NA
2-A TD

FIG. 7

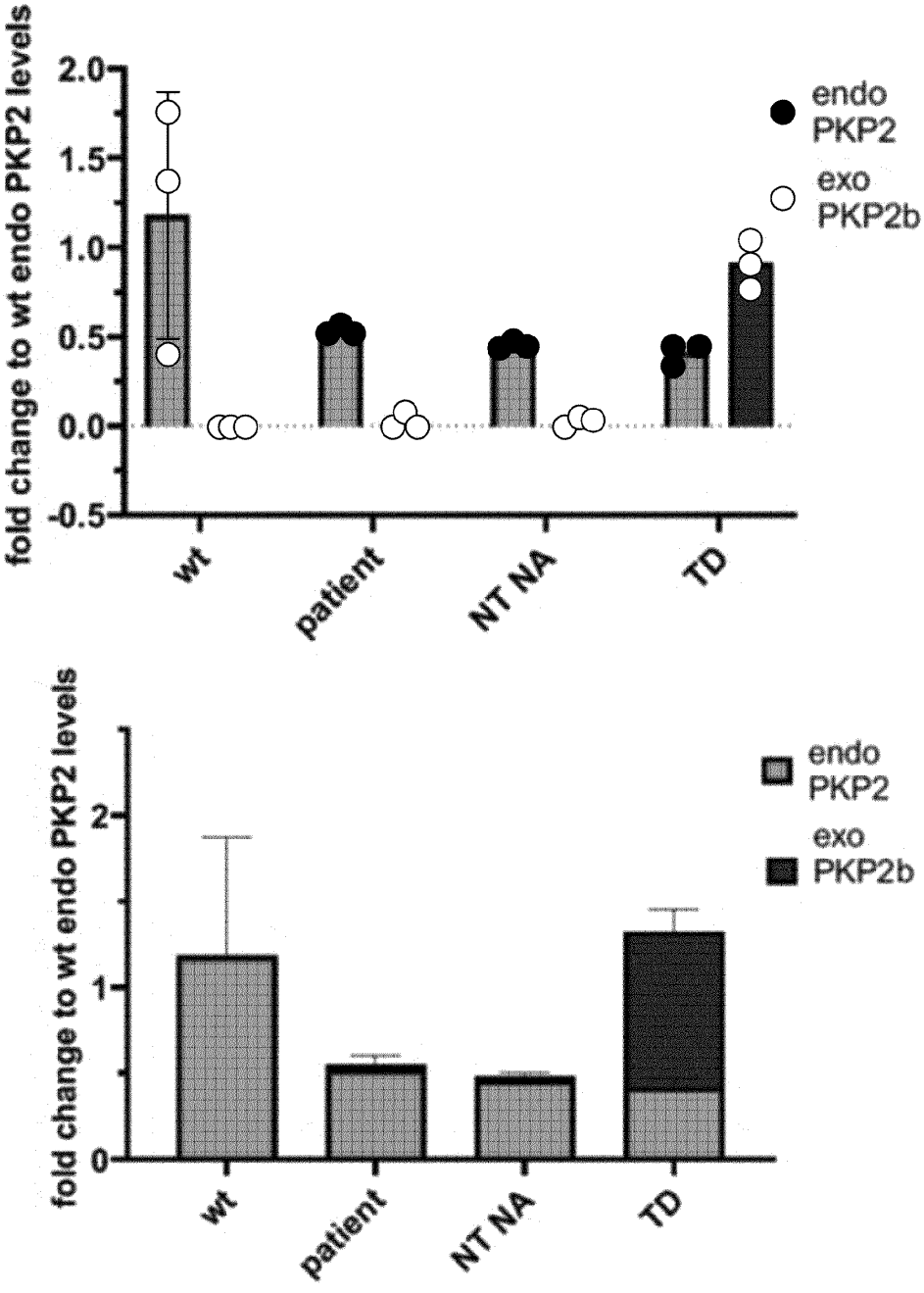
fold change to wt endo PKP2 levels
2.0
1.5
1.0
0.5
0.0
-0.5
endo PKP2
exo PKP2b
wt
patient
NT NA
TD
fold change to wt endo PKP2 levels
2
1
0
endo PKP2
exo PKP2b
wt
patient
NT NA
TD

FIG. 8

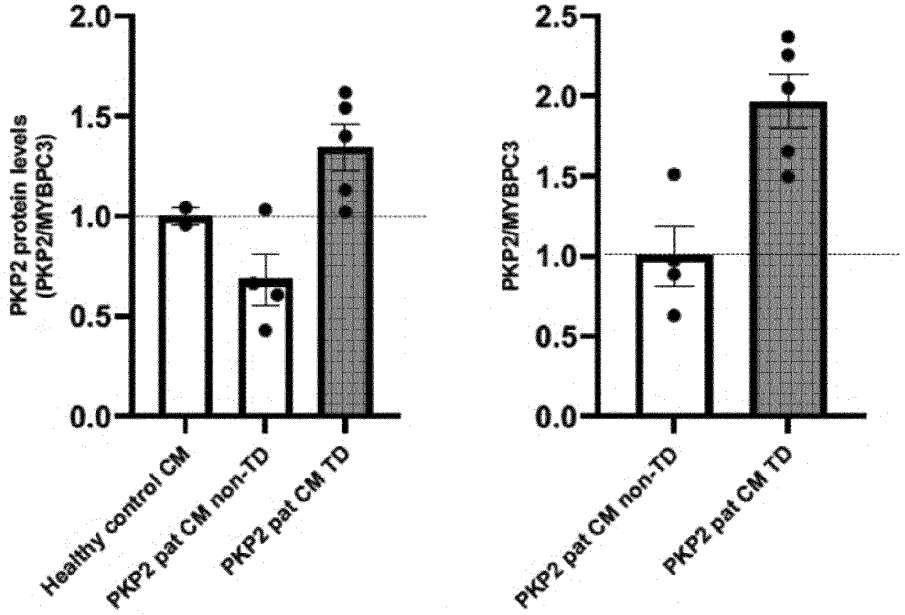
PKP2 protein levels (PKP2/MYBPC3)
2.0
1.5
1.0
0.5
0.0
Healthy control CM
PKP2 pat CM non-TD
PKP2 pat CM TD
PKP2/MYBPC3
2.5
2.0
1.5
1.0
0.5
0.0
PKP2 pat CM non-TD
PKP2 pat CM TD

FIG. 9A

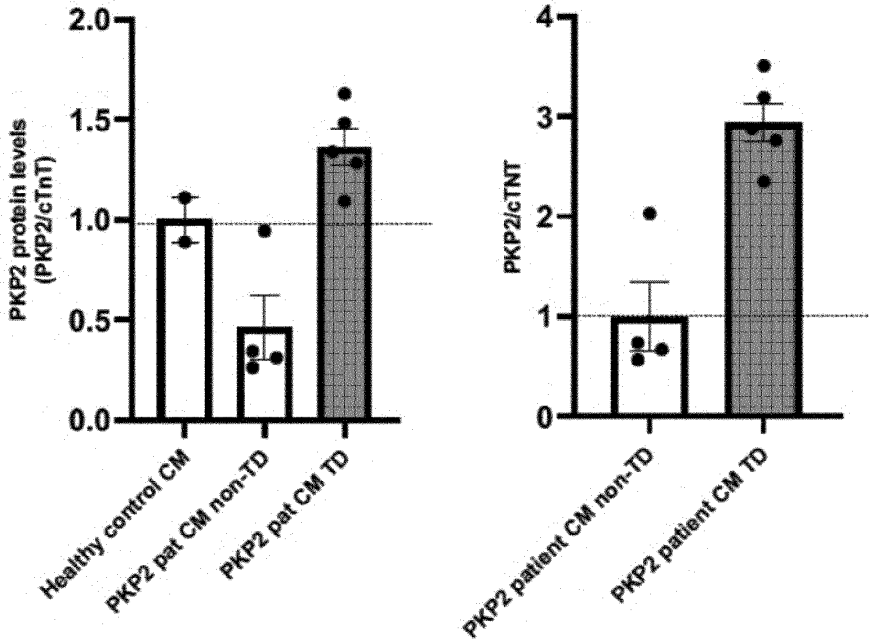
PKP2 protein levels (PKP2/cTnT)
2.0
1.5
1.0
0.5
0.0
Healthy control CM
PKP2 pat CM non-TD
PKP2 pat CM TD
PKP2/cTNT
4
3
2
1
0
PKP2 patient CM non-TD
PKP2 patient CM TD

FIG. 9B

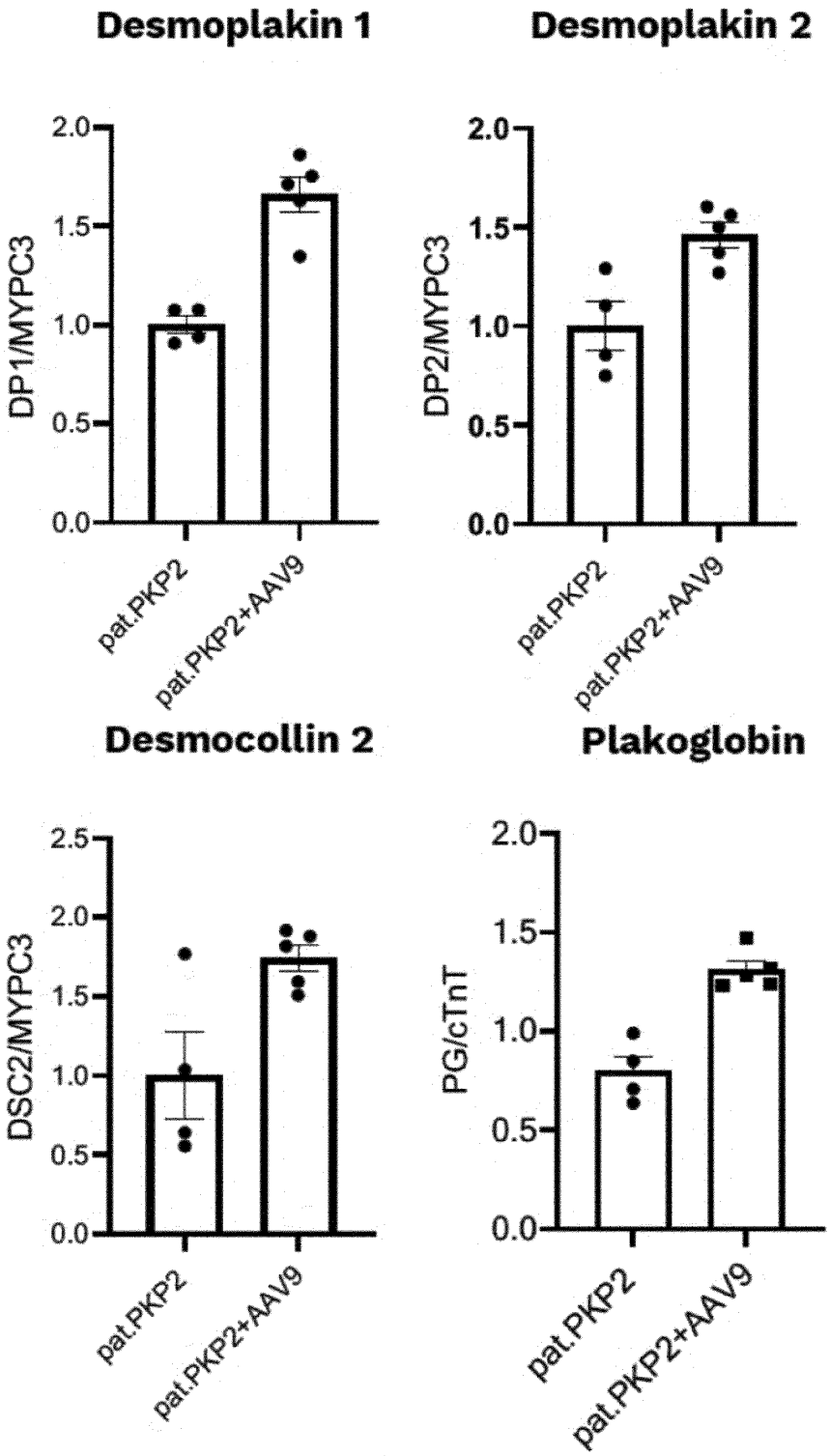
Desmoplakin 1
DP1/MYPC3
2.0
1.5
1.0
0.5
0.0
pat.PKP2
pat.PKP2+AAV9
Desmoplakin 2
DP2/MYPC3
2.0
1.5
1.0
0.5
0.0
pat.PKP2
pat.PKP2+AAV9
Desmocollin 2
DSC2/MYPC3
2.5
2.0
1.5
1.0
0.5
0.0
pat.PKP2
pat.PKP2+AAV9
Plakoglobin
PG/cTnT
2.0
1.5
1.0
0.5
0.0
pat.PKP2
pat.PKP2+AAV9

FIG. 10A

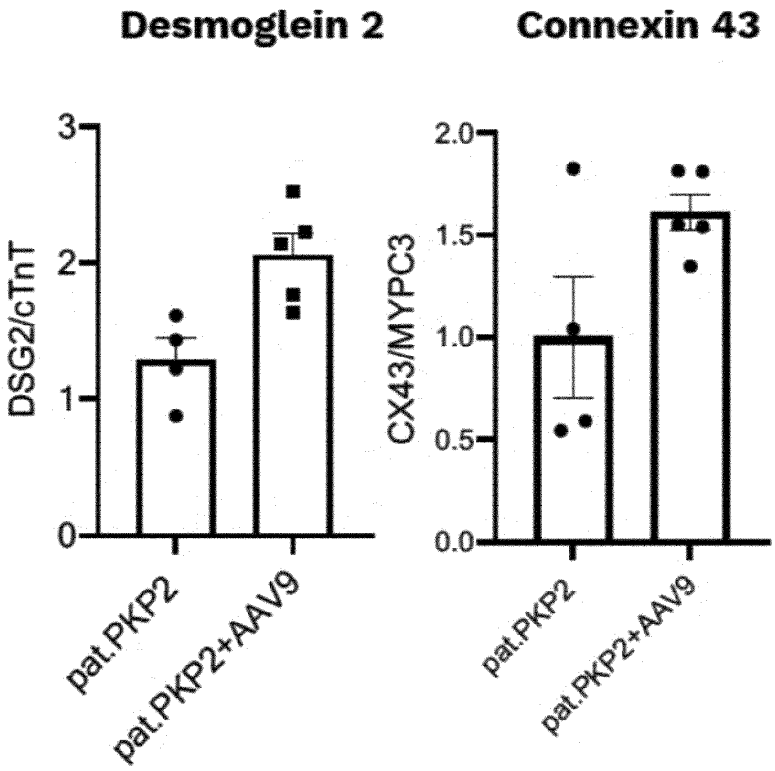
Desmoglein 2
Connexin 43
DSG2/cTnT
CX43/MYPC3
3
2
1
0
2.0
1.5
1.0
0.5
0.0
pat.PKP2
pat.PKP2+AAV9
pat.PKP2
pat.PKP2+AAV9

FIG. 10B