

Supplementary figure and AVI legends.

Supplementary Fig. 1. Near-unique example of mucus secretion from submucosal glands from a CF airway in response to 10 SubP + 1 μ M phosphoramidon and Forskolin. This CF subject (homozygous DF508) was unique in responding to both SubP and the combination of SubP + forskolin (2 of 6 glands responded to SubP and an additional 2 glands responded to the combined stimuli). One possible explanation for these responses is that this was the freshest CF tissue tested (2 hr post harvest).

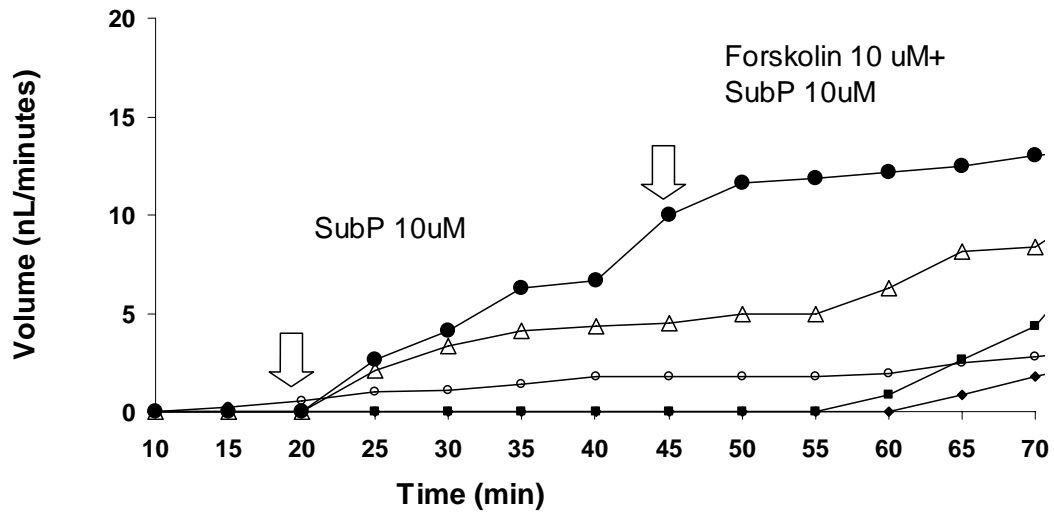
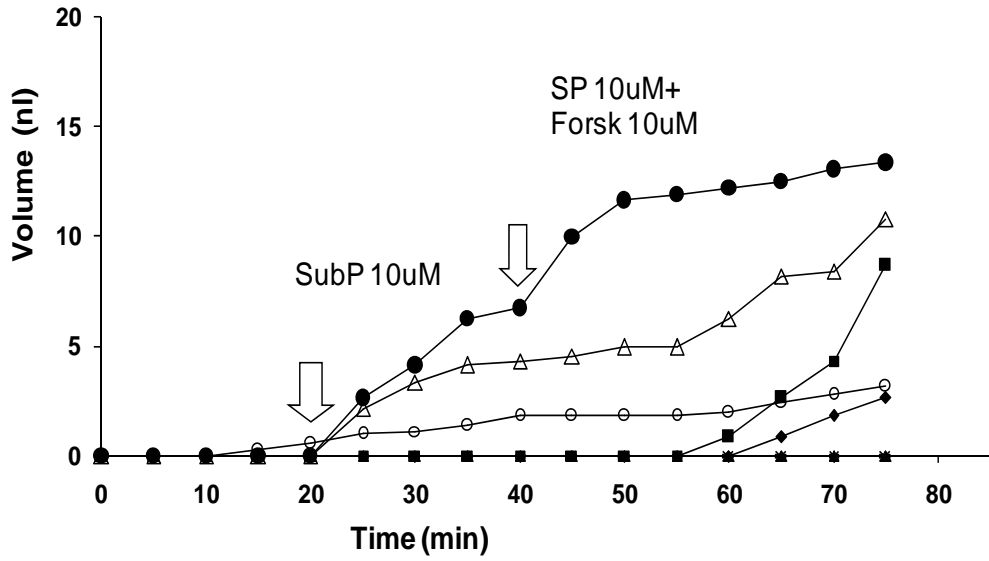
Supplementary Fig. 2. Proportion of airway glands responding in the synergy paradigm to VIP or forskolin alone or with SubP, as a function of the total number of glands that responded to a final addition of 10 μ M carbachol. A. Donor control subjects; B. Disease control subjects; C. Cystic Fibrosis subjects.

Supplementary AVI-01: HN152 Gland from donor trachea. Secretion from multiple acini to 10 μ M substance P added in the presence of 1 μ M phosphoramidon which was added 10 min before the SubP; 10 sec intervals. Note initial contraction of myoepithelial cells and then subsequent expansion caused by fluid secretion: particulates in mucus allow flow to be observed clearly in this preparation; note slowing of flow with continued presence of SubP.

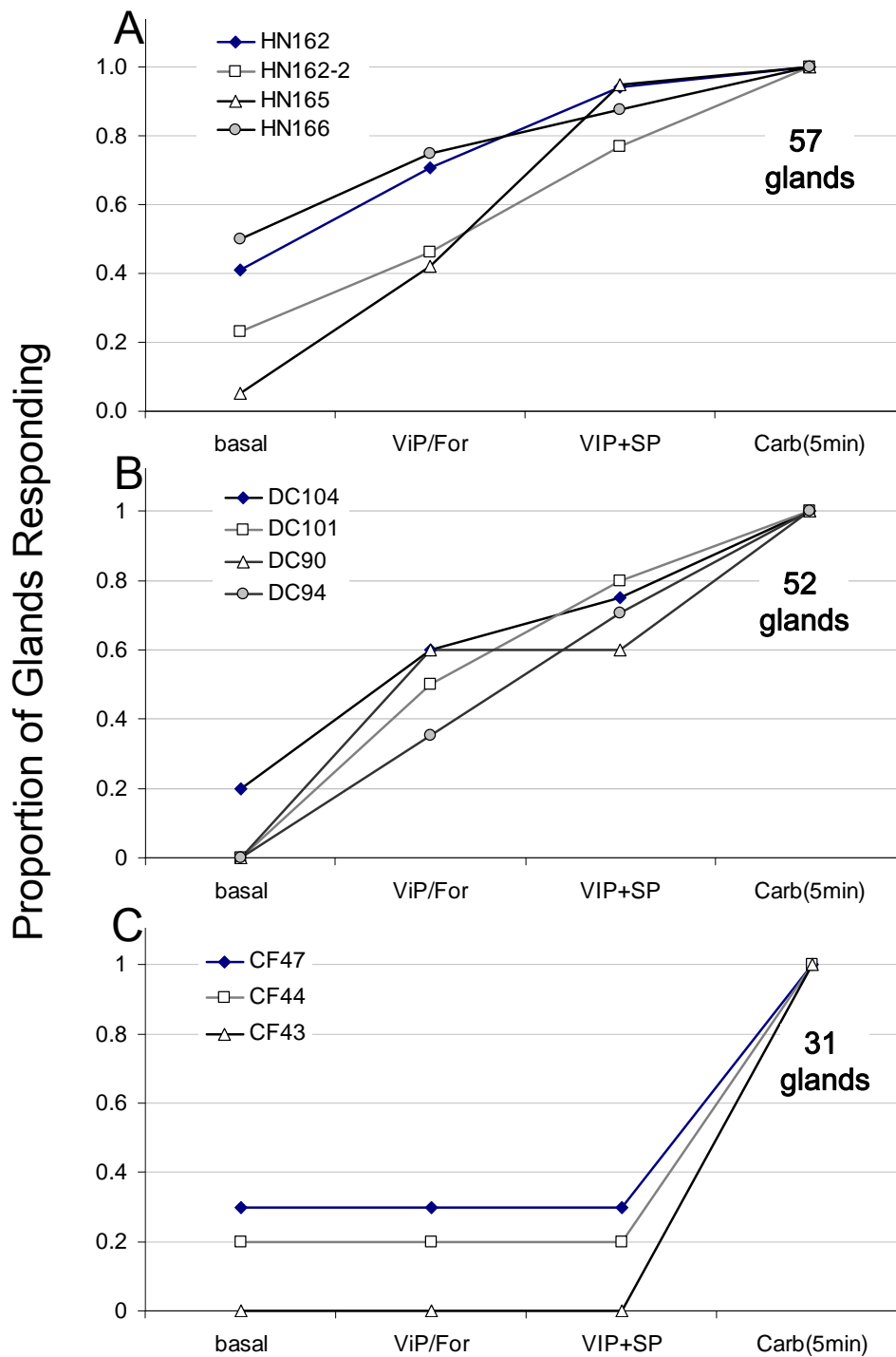
Supplementary AVI-2: DC100 Gland from bronchus of COPD patient. Secretion from multiple acini in response to 10 μ M SubP + 1 μ M phosphoramidon added at frame 48. 10 sec intervals. Sheets of mucus can be seen flowing in the larger tubule.

Supplementary AVI-3: CF40 Response to 1 μ M SubP in Krebs (without phosphoramidon) shows strong but brief myoepithelial cell contraction with some expulsion of luminal contents, but with decrease rather than increase in lumen volume and little evidence of cell volume loss or fluid secretion.

Supplementary Fig 1



Supplementary Fig 2



Supplementary Fig. 2. Proportion of airway glands responding in the synergy paradigm to VIP or forskolin alone or with SubP, as a function of the total number of glands that responded to a final addition of 10 μ M carbachol. A. Donor control subjects; B. Disease control subjects; C. Cystic Fibrosis subjects.