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## References

- Gupta N, Vassallo R, Wikenheiser-Brokamp KA, McCormack FX. Diffuse cystic lung disease: part I. *Am J Respir Crit Care Med* 2015; 191:1354–1366.
- Gupta R, Kitaichi M, Inoue Y, Kotloff R, McCormack FX. Lymphatic manifestations of lymphangioleiomyomatosis. *Lymphology* 2014;47: 106–117.
- Kumasaka T, Seyama K, Mitani K, Sato T, Souma S, Kondo T, et al. Lymphangiogenesis in lymphangioleiomyomatosis: its implication in the progression of lymphangioleiomyomatosis. *Am J Surg Pathol* 2004;28:1007–1016.
- Kumasaka T, Seyama K, Mitani K, Souma S, Kashiwagi S, Hebisawa A, et al. Lymphangiogenesis-mediated shedding of LAM cell clusters as a mechanism for dissemination in lymphangioleiomyomatosis. *Am J Surg Pathol* 2005;29:1356–1366.
- Gupta N, Lee HS, Young LR, Strange C, Moss J, Singer LG, et al.; NIH Rare Lung Disease Consortium. Analysis of the MILES cohort reveals determinants of disease progression and treatment response in lymphangioleiomyomatosis. *Eur Respir J* 2019;53:1802066.
- McCormack FX, Gupta N, Finlay GR, Young LR, Taveira-DaSilva AM, Glasgow CG, et al.; ATS/JRS Committee on Lymphangioleiomyomatosis. Official American Thoracic Society/Japanese Respiratory Society clinical practice guidelines: lymphangioleiomyomatosis diagnosis and management. *Am J Respir Crit Care Med* 2016;194:748–761.
- Young L, Lee HS, Inoue Y, Moss J, Singer LG, Strange C, et al.; MILES Trial Group. Serum VEGF-D a concentration as a biomarker of lymphangioleiomyomatosis severity and treatment response: a prospective analysis of the Multicenter International Lymphangioleiomyomatosis Efficacy of Sirolimus (MILES) trial. *Lancet Respir Med* 2013;1:445–452.
- Young LR, Inoue Y, McCormack FX. Diagnostic potential of serum VEGF-D for lymphangioleiomyomatosis. *N Engl J Med* 2008;358:199–200.
- Young LR, Vandyke R, Gulleman PM, Inoue Y, Brown KK, Schmidt LS, et al. Serum vascular endothelial growth factor-D prospectively distinguishes lymphangioleiomyomatosis from other diseases. *Chest* 2010;138:674–681.
- McCormack FX, Inoue Y, Moss J, Singer LG, Strange C, Nakata K, et al.; National Institutes of Health Rare Lung Diseases Consortium; MILES Trial Group. Efficacy and safety of sirolimus in lymphangioleiomyomatosis. *N Engl J Med* 2011;364:1595–1606.
- Seyama K, Kumasaka T, Souma S, Sato T, Kurihara M, Mitani K, et al. Vascular endothelial growth factor-D is increased in serum of patients with lymphangioleiomyomatosis. *Lymphat Res Biol* 2006;4:143–152.
- Wartiovaara U, Salven P, Mikkola H, Lassila R, Kaukonen J, Joukov V, et al. Peripheral blood platelets express VEGF-C and VEGF which are released during platelet activation. *Thromb Haemost* 1998;80:171–175.

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## Peer Support Group for Intensive Care Unit Survivors: Perceptions on Supportive Recovery in the Era of Social Distancing

To the Editor:

Up to 70% of intensive care unit (ICU) survivors experience long-term cognitive impairment, psychological difficulties, and

physical disability (1–5). Referred to as “post-intensive care syndrome” (PICS), this collection of symptoms presents challenges to survivors and caregivers that limit functional independence, employment, and quality of life (6–10). Despite their prevalence, symptoms are frequently underrecognized and undertreated (5, 11). With an estimated 5.7 million ICU admissions annually in the United States (12), expected to increase alongside the evolving coronavirus disease 2019 (COVID-19) pandemic (13), the focus on recovery after critical illness is a major public health concern that demands adaptive strategies to facilitate recovery for this vulnerable population.

In-person (14, 15) and virtual (16–21) peer support groups help overcome barriers to PICS recovery, such as limited patient education on PICS and access to post-acute care psychosocial interventions. Therapeutic support groups unite people facing similar issues, emphasize emotional support, and incorporate educational components. Virtual support systems for chronic disease management have great potential during the COVID-19 pandemic. Despite a paucity of data for psychosocial interventions for ICU survivors, we must disseminate best practices to address the anticipated needs of COVID-19 survivors.

The ICU Survivor Peer Support Group at the Critical Illness, Brain Dysfunction, and Survivorship Center has met weekly for 10 years, providing structured and informal support for a growing number of ICU survivors across the United States. With new restrictions and concerns during the COVID-19 pandemic, we

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transitioned our hybrid (in-person and virtual) support group to an entirely virtual format (22). We aim to describe the value of our group for post-ICU recovery and discuss the feasibility and acceptability of a fully virtual support group.

### Methods

After local institutional-review-board approval, we conducted an anonymous online survey of our ICU Survivor Peer Support Group participants and facilitators 6 weeks after transitioning to the virtual group. Eligible members ( $N=84$ ) were obtained from a support group listserv and were given 2 weeks for completion of the survey. Survey components were designed by study investigators. Demographic data, including sex, age range, and distance from the host institution, were obtained by voluntary response. Value, feasibility, and barriers to the support group were collected by using a Likert scale, multiple-choice questions, and free text. Data were collected and managed using Research Electronic Data Capture, which is hosted at Vanderbilt University (23).

**Statistical analysis.** Descriptive statistics were analyzed with Stata version 16 (StataCorp LLC) (24).

### Results

The survey response rate was 31.0% (26 of 84 participants) and included responses from 22 group participants and 4 facilitator respondents. Although we sent the survey to 78 participants and 6 facilitators, the 26 respondents represented mostly *regular* group attendees. Demographics are presented in Table 1.

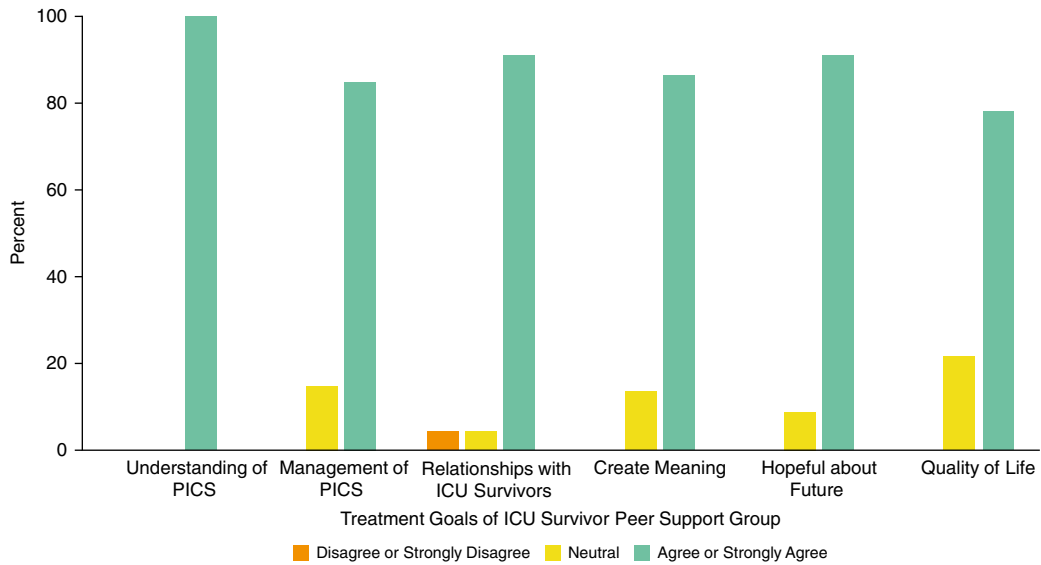
**Value of the peer support group.** The majority of participant respondents “agreed” or “strongly agreed” that the peer support group helped them to gain a greater understanding of PICS, better manage symptoms of PICS, and create relationships with others who had experienced ICU hospitalizations (Figure 1). Participants believed that the support group helped them to create meaning from their ICU experiences and feel more hopeful about the future and that it improved their quality of life.

**Feasibility of the virtual peer support group.** Before adopting the entirely virtual format, 69.0% (18 of 26) of respondents participated in person. To attend the virtual peer support group, participants used a mobile phone (with or without video), tablet, or computer. The majority of participants (92.0%; 24 of 26) felt comfortable or very comfortable using their device to attend the

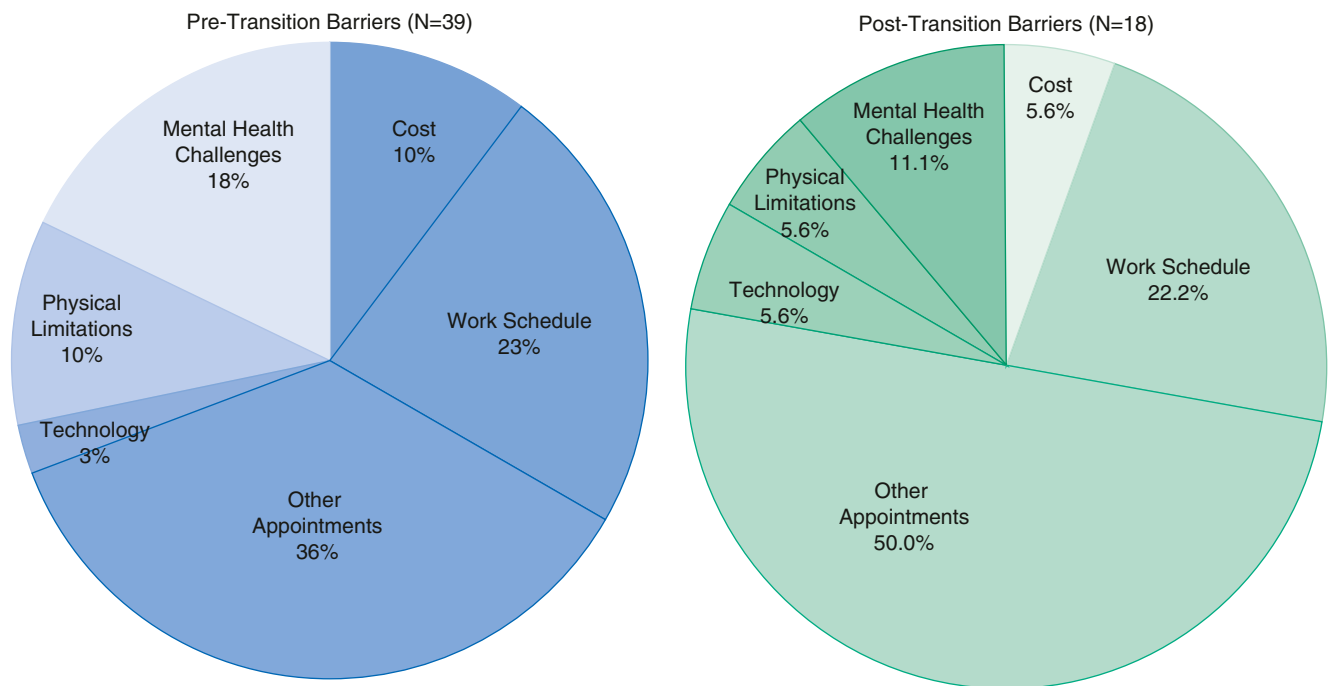
**Table 1.** Survey-respondent demographics

Characteristic	Patients [n (%)] (N=20)	Caregivers [n (%)] (N=2)	Facilitators [n (%)] (N=4)
Age			
18–29 yr	—	—	2 (50)
30–39 yr	3 (15)	1 (50)	2 (50)
50–59 yr	6 (30)	—	—
60–69 yr	8 (40)	—	—
70–79 yr	3 (15)	1 (50)	—
Sex			
Female	6 (30)	1 (50)	4 (100)
Male	14 (70)	—	—
Unavailable	—	1 (50)	—
Education			
12th-grade level or lower	—	—	—
High school diploma/GED	1 (5)	—	—
Some college	2 (10)	—	—
Associate’s degree	6 (30)	—	—
Bachelor’s degree	6 (30)	2 (100)	—
Master’s degree	2 (10)	—	1 (25)
Doctoral degree	3 (15)	—	3 (100)
Distance from Vanderbilt			
0–20 miles	7 (35)	1 (50)	4 (100)
21–50 miles	6 (30)	1 (50)	—
51–100 miles	—	—	—
>100 miles	7 (35)	—	—
Time since most recent ICU hospitalization			
<6 mo	3 (15)	—	—
6 mo to 1 yr	2 (10)	—	—
1–2 yr	7 (35)	—	—
3–4 yr	1 (5)	—	—
5–10 yr	6 (30)	—	—
>10 yr	1 (5)	—	—
Groups attended in last year			
<5	3 (15)	—	—
≥5	17 (85)	2 (100)	4 (100)
Frequency of attendance			
<1 time/mo	—	1 (50)	—
1–2 groups/mo	2 (13.3)	1 (50)	—
3–4 groups/mo	13 (86.7)	—	4 (100)

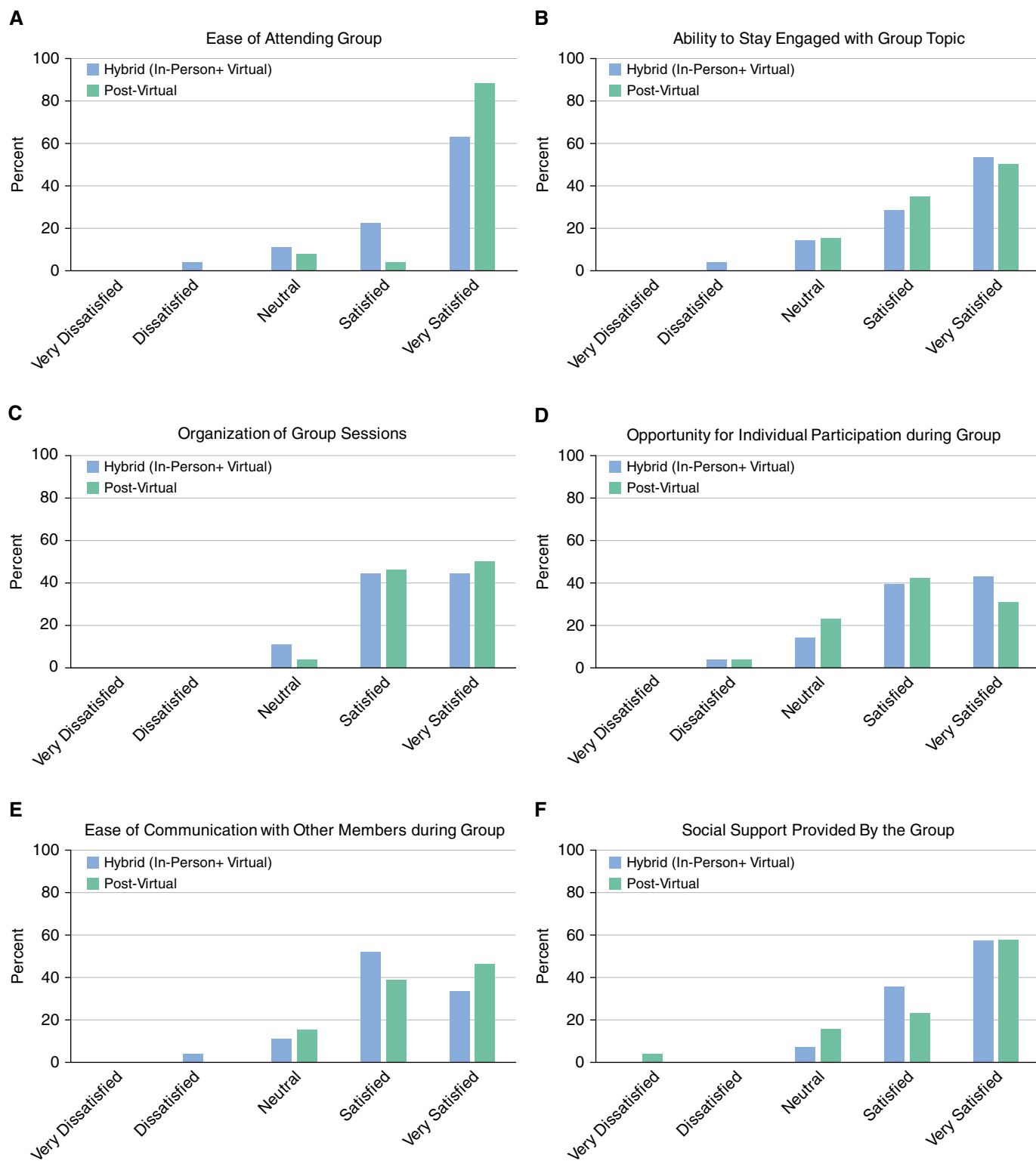
Definition of abbreviations: GED=general education diploma; ICU=intensive care unit.



**Figure 1.** Participant responses ( $n = 22$ ) to value-assessment statements were obtained using the Likert scale (options included “strongly disagree,” “disagree,” “neutral,” “agree,” and “strongly agree”). Participants include intensive care unit (ICU) survivors and their caregivers. Statements were constructed by authors to assess for the value of the ICU peer support group and included the following six statements, respectively: “The support group has helped me gain a greater understanding of PICS,” “The support group has helped me learn how to better manage my symptoms of PICS,” “The support group has helped me create relationships with others who had similar experiences during and after ICU hospitalization,” “The support group has helped me create meaning from my experiences related to ICU hospitalization,” “I feel more hopeful about my future by attending the support group,” and “My quality of life has improved as a result of the support group.” PICS = post-intensive care syndrome.



**Figure 2.** Twenty-two (84.6%) survey respondents endorsed at least one barrier to group attendance. Participants were asked to endorse perceived barriers to attendance of at least two support group sessions before and after the transition to a full virtual support group for intensive care unit survivors. Participants endorsed a total of 34 and 18 barriers to attendance of the hybrid and virtual support groups, respectively. Categories are not mutually exclusive. Endorsement of barriers to attendance of both the hybrid group and virtual group was completed retrospectively via survey 6–8 weeks after the transition to the fully virtual group format.



**Figure 3.** Participant-satisfaction responses were obtained from participants (intensive care unit survivors and caretakers;  $n = 22$ ) to determine the impact of transition from a hybrid (in-person + virtual) to a virtual peer support group format. Questions were obtained on a Likert scale (options included “strongly disagree,” “disagree,” “neutral,” “agree,” and “strongly agree”). Responses assessed core support group components, including (A) ease of attendance, (B) ability to stay engaged, (C) session organization, (D) opportunity for participation, (E) ease of communication, and (F) social support. Participant-satisfaction ratings of the hybrid (in-person + virtual) group are based on retrospective ratings completed 6–8 weeks after transition to the fully virtual format.

group. Participants endorsed fewer barriers to attendance after transition to the virtual group (Figure 2).

**Satisfaction with virtual peer support group.** Participant-satisfaction ratings for the virtual support group are shown in Figure 3. After transition to the virtual format, 90.0% (20 of 22) of participants stated they were likely or very likely to continue to attend the virtual group.

## Discussion

This brief assessment of our ICU Survivors Peer Support Group suggests that virtual peer support groups for ICU survivors are valuable, feasible, and acceptable, with patient-reported benefits including social connection and support, greater understanding of physical and emotional health, increased knowledge of illness management, and improved quality of life. There were high rates of satisfaction on comfort with technology and all measures of acceptability. Participant-satisfaction ratings for the ease of attending the group improved after the transition to the virtual format. Overall, participants reported satisfaction with the virtual support group, suggesting that this format is an acceptable alternative, potentially mitigating barriers to mental health support for ICU survivors. Although our group did not include survivors of COVID-19 who were admitted to the ICU, group benefits may be critical to these patients, who face isolation, decreased social support, economic hardship, and health-related anxiety as part of this rapidly evolving global pandemic.

Improved access to psychosocial support during an era with increasing mental health risk factors will be a necessary component of post-ICU recovery. Despite concerns that virtual groups may foster feelings of disconnection (16, 25), our group participants reported a greater degree of satisfaction with the social support provided in the all-virtual group than they reported with the hybrid group.

This study has several limitations, including its small sample size, study design and response bias, and limited generalizability. Our group uniquely incorporated virtual attendees before COVID-19. Although many in-person participants established social relationships with one another before using a virtual platform, which may have facilitated the successful transition, almost one-third of respondents never participated in person but nevertheless found the group valuable. Group members may not necessarily represent the emerging population of ICU survivors whose ICU admission was related to COVID-19, who could benefit from virtual support groups because of unifying issues of survivorship across primary diagnoses (26, 27). Our experience suggests that virtual support groups are valuable and feasible for an expanding population of ICU survivors.

Future considerations for sustaining virtual peer support groups include confidentiality, optimizing virtual therapeutic relationships, pregroup screening, and significant efforts to address health inequities and disparities that would otherwise leave vulnerable populations without access to virtual mental health services.

## Conclusions

We share this study with *urgency* to increase awareness and encourage use of virtual support groups for ICU survivors. As the COVID-19 pandemic evolves, we have the responsibility to provide safe and accessible health care for ICU survivors. Undoubtedly, social isolation presents a risk to the mental health and quality of life of survivors. Virtual platforms for peer support groups are one way we can increase access to social support, maintain human

connection, and lessen the mounting psychological distress experienced by survivors of critical illness.

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## References

- Pandharipande PP, Girard TD, Jackson JC, Morandi A, Thompson JL, Pun BT, *et al.*; BRAIN-ICU Study Investigators. Long-term cognitive impairment after critical illness. *N Engl J Med* 2013;369:1306–1316.
- Girard TD, Jackson JC, Pandharipande PP, Pun BT, Thompson JL, Shintani AK, *et al.* Delirium as a predictor of long-term cognitive impairment in survivors of critical illness. *Crit Care Med* 2010;38:1513–1520.
- Jackson JC, Pandharipande PP, Girard TD, Brummel NE, Thompson JL, Hughes CG, *et al.*; Bringing to Light the Risk Factors and Incidence of Neuropsychological Dysfunction in ICU Survivors (BRAIN-ICU) Study Investigators. Depression, post-traumatic stress disorder, and functional disability in survivors of critical illness in the BRAIN-ICU study: a longitudinal cohort study. *Lancet Respir Med* 2014;2:369–379.
- Jackson JCH, Hart RP, Gordon SM, Shintani A, Truman B, May L, *et al.* Six-month neuropsychological outcome of medical intensive care unit patients. *Crit Care Med* 2003;31:1226–1234.
- Desai SV, Law TJ, Needham DM. Long-term complications of critical care. *Crit Care Med* 2011;39:371–379.
- Hatch R, Young D, Barber V, Griffiths J, Harrison DA, Watkinson P. Anxiety, depression and post traumatic stress disorder after critical illness: a UK-wide prospective cohort study. *Crit Care* 2018;22:310.
- Davidson JE, Jones C, Bienvenu OJ. Family response to critical illness: postintensive care syndrome—family. *Crit Care Med* 2012;40:618–624.
- Mikkelsen ME, Christie JD, Lanke PN, Biester RC, Thompson BT, Bellamy SL, *et al.* The adult Respiratory Distress Syndrome Cognitive Outcomes Study: long-term neuropsychological function in survivors of acute lung injury. *Am J Respir Crit Care Med* 2012;185:1307–1315.
- Hopkins RO, Weaver LK, Collingridge D, Parkinson RB, Chan KJ, Orme JF Jr. Two-year cognitive, emotional, and quality-of-life outcomes in acute respiratory distress syndrome. *Am J Respir Crit Care Med* 2005;171:340–347.
- Rawal G, Yadav S, Kumar R. Post-intensive care syndrome: an overview. *J Transl Int Med* 2017;5:90–92.
- Myers EA, Smith DA, Allen SR, Kaplan LJ. Post-ICU syndrome: rescuing the undiagnosed. *JAAPA* 2016;29:34–37.
- Halpern NA, Pastores SM. Critical care medicine in the United States 2000–2005: an analysis of bed numbers, occupancy rates, payer mix, and costs. *Crit Care Med* 2010;38:65–71.
- Cook DJ, Marshall JC, Fowler RA. Critical illness in patients with COVID-19: mounting an effective clinical and research response. *JAMA* 2020;323:1559–1560.

- 14 Daniels LM, Johnson AB, Cornelius PJ, Bowron C, Lehnertz A, Moore M, et al. Improving quality of life in patients at risk for post-intensive care syndrome. *Mayo Clin Proc Innov Qual Outcomes* 2018;2:359–369.
- 15 Vlasek JH, van Genderen ME, Schut A, Verkade M, Wils EJ, Gommers D, et al. Patients suffering from psychological impairments following critical illness are in need of information. *J Intensive Care* 2020;8:6.
- 16 Flickinger TE, DeBolt C, Waldman AL, Reynolds G, Cohn WF, Beach MC, et al. Social support in a virtual community: analysis of a clinic-affiliated online support group for persons living with HIV/AIDS. *AIDS Behav* 2017;21:3087–3099.
- 17 Armstrong MJ, Alliance S. Virtual support groups for informal caregivers of individuals with dementia: a scoping Review. *Alzheimer Dis Assoc Disord* 2019;33:362–369.
- 18 Stewart MJ, Hart G, Mann K, Jackson S, Langille L, Reidy M. Telephone support group intervention for persons with hemophilia and HIV/AIDS and family caregivers. *Int J Nurs Stud* 2001;38:209–225.
- 19 Dale J, Caramlau IO, Lindenmeyer A, Williams SM. Peer support telephone calls for improving health. *Cochrane Database Syst Rev* 2008;2008:CD006903.
- 20 Smith TL, Toseland RW. The effectiveness of a telephone support program for caregivers of frail older adults. *Gerontologist* 2006;46:620–629.
- 21 Banbury A, Nancarrow S, Dart J, Gray L, Parkinson L. Telehealth interventions delivering home-based support group videoconferencing: systematic Review. *J Med Internet Res* 2018;20:e25.
- 22 Zoom Video Communications, Inc. Security guide. San Jose, CA: Zoom Video Communications; 2020 [accessed 2020 Jun 24]. Available from: <https://zoom.us/docs/doc/Zoom-Security-White-Paper.pdf>.
- 23 Harris PA, Taylor R, Thielke R, Payne J, Gonzalez N, Conde JG. Research Electronic Data Capture (REDCap): a metadata-driven methodology and workflow process for providing translational research informatics support. *J Biomed Inform* 2009;42:377–381.
- 24 StataCorp. Stata statistical software: release 16. College Station, TX: StataCorp; 2019.
- 25 Kozlowski KA, Holmes CM. Experiences in online process groups: a qualitative study. *J Spec Group Work* 2014;39:276–300.
- 26 Hosey MM, Needham DM. Survivorship after COVID-19 ICU stay. *Nat Rev Dis Primers* 2020;6:60.
- 27 Korupolu R, Francisco GE, Levin H, Needham DM. Rehabilitation of critically ill COVID-19 survivors. *J Int Soc Phys Rehabil Med* 2020;3:45–52.

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## Appearance of Pancreatic Sufficiency and Discontinuation of Pancreatic Enzyme Replacement Therapy in Children with Cystic Fibrosis on Ivacaftor

To the Editor:

In their paper examining the safety and efficacy of ivacaftor in children aged 2–5 years in 2016, Davies and colleagues (1) describe an increase in fecal elastase (FE) in a group of 27 children over 24 weeks in a phase 3 trial. Sustained improvements were seen in the open-label study over 84 weeks in this cohort (2). Isolated case reports have confirmed increases in FE levels on treatment and, in two cases, cessation of pancreatic enzyme replacement therapy (PERT) treatment (3–5). Nichols and colleagues recently reported that 7 of 17 children converted from pancreatic insufficiency to sufficiency on ivacaftor over a mean period of 5 years, with two discontinuing PERT altogether (6). On the basis of early trial data, we have been monitoring FE and clinical symptoms in children on ivacaftor at our center and instituting a carefully monitored trial of discontinuing PERT when FE has normalized.

Eighteen children at our cystic fibrosis (CF) center with G551D CFTR mutations have started ivacaftor in the last 11 years at a mean age of 5.8 years (range 1–11.4 yr) (Figure 1). The mean duration of treatment is 4.5 years (range 0.6–11.4 yr). FE levels have increased in all but one individual, with 11 returning to the pancreatic sufficiency range (>200 mcg/g), three to the 100–200 mcg/g range, and three becoming detectable. Children achieving sufficiency were more likely to have had detectable FE at baseline (8/11 vs. 0/7,  $P < 0.01$ ), less likely to have a second “severe” CF mutation (F508del or minimum function; 2/11 vs. 6/7;  $P = 0.01$ ), and more likely to be younger at ivacaftor commencement (mean 4.0 yr vs. 8.6 yr;  $P < 0.001$ ). All 11 sufficient children have discontinued PERT without the development of abdominal pain, weight loss, or steatorrhea. Symptoms are monitored using a standard clinical proforma delivered by a single dietician at three monthly intervals in all children. The median follow-up after discontinuation is 12 months (8–22 mo). PERT usage reduced in all but one individual after starting ivacaftor. In the seven individuals who have not discontinued ivacaftor, parent-reported PERT usage has significantly reduced (mean, 5,302 vs.

3,509 U/kg;  $P = 0.01$ ) without the development of abdominal pain, weight loss, or steatorrhea in any subject.

This is a retrospective review of clinically collected data, so it lacks the rigor of a prospective study. With a relatively short follow-up, we cannot say definitively yet that discontinuation in our subjects is without consequence; however clinical experience with pancreatic insufficiency is that discontinuation of PERT will usually result in obvious symptoms within days and adverse nutritional consequences within weeks or months. Ongoing annual surveillance of FE levels in all individuals in whom PERT has been discontinued is underway. Of particular note are the children with undetectable levels of FE, two of whom (starting at age 6 yr) reached sufficiency after 3–5 years, and two (starting at ages 8 and 9) who almost achieved sufficiency after 7 years. These latter two subjects have unilaterally discontinued enzymes before measurement of FE levels on the basis of the lack of symptoms. FE levels vary on treatment between individuals and in individuals between measures. Several children have had moved between pancreatic insufficiency and pancreatic sufficiency categories in both directions, suggesting that a return to insufficiency may be possible in some on treatment. The slow pace of recovery here is intriguing and is consistent with previous reports but incongruent with the effects seen in other organs, including the lungs, bowel and sweat glands.

Our data show that improvements in FE, even after several years, can reflect clinical pancreatic sufficiency and suggest that we should closely monitor FE levels, symptoms, and enzyme use for many years after modulator treatment begins. The variability in FE levels and concerns about return of insufficiency suggest that FE and symptoms should be serially measured after the discontinuation of PERT.

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