NICE National Institute for Health and Care Excellence

Surveillance report 2017 – Idiopathic pulmonary fibrosis in adults: diagnosis and management (2013) NICE guideline CG163

Surveillance report

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Contents

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Surveillance decision

We will not update the guideline on idiopathic pulmonary fibrosis (IPF) at this time.

We will amend the guideline to:

- Add a cross-reference to NICE technology appraisal guidance on nintedanib for treating idiopathic pulmonary fibrosis, published in January 2016, from recommendations 1.5.11–1.5.15.

Reason for the decision

Assessing the evidence

We found 38 studies through surveillance of this guideline.

We identified evidence that supports current recommendations on:

- diagnosis – multidisciplinary team
- information and support
- management – pulmonary rehabilitation
- management – disease-modifying pharmacological interventions.

We also found new evidence that was not thought to have an impact on current recommendations, including:

- management – best supportive care.

We asked topic experts whether this new evidence would affect current recommendations on diagnosis and management of IPF in this guideline. Generally, the topic experts thought that an update was not needed.

We did not find any evidence in areas not covered by the original guideline.

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We did not find any evidence related to:

- awareness of clinical features of IPF
- prognosis
- management – ventilation
- review and follow-up.

For any new evidence relating to published or ongoing NICE technology appraisals, the guideline surveillance review deferred to the technology appraisal decision. This included nintedanib for treating idiopathic pulmonary fibrosis, published in January 2016.

Equalities

No equalities issues were identified during the surveillance process.

Overall decision

After considering all the evidence and views of topic experts and stakeholders, we decided that no update is necessary for this guideline.

See how we made the decision for further information.

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Commentary on selected evidence

With advice from topic experts we selected 2 studies for further commentary.

Information and support

We selected a qualitative study, Sampson et al. (2015), for a full commentary because it is a UK-based study that emphasises the need for individualised care and psychosocial support at key transition points for patients diagnosed with idiopathic pulmonary fibrosis (IPF) and their carers.

What the guideline recommends

NICE guideline CG163 recommends that the consultant respiratory physician or interstitial lung disease specialist nurse should provide accurate and clear information (verbal and written) to people with IPF, and their families and carers. This should include information about investigations, diagnosis and management, and an interstitial lung disease specialist nurse should be available at all stages of the care pathway to provide information and support to people with IPF, and their families and carers (recommendations 1.3.1–1.3.4).

Methods

Sampson et al. (2015) conducted a qualitative study aiming to explore the perspectives of patients and their carers across the IPF spectrum to inform the development of clinical pathways and multidisciplinary service interventions.

Recruitment was carried out at 2 UK specialist interstitial lung disease clinics. Twenty seven patients (18 men and 9 women) with IPF and 21 carers (6 men and 15 women) were included. Six patients had no carer and none of the patients had been prescribed pirfenidone. A carer was defined as a person who provided emotional support and contributed most to the patient's care in the earlier stages of disease. Patients' IPF was categorised according to disease extent. An experienced qualitative researcher carried out semi-structured interviews with participants. Anonymised transcripts were analysed by the same researcher using Interpretative Phenomenological Analysis (IPA). The generated themes from the analysis were discussed and agreed by other members of the research team. The consolidated criterion for reporting qualitative research (COREQ) 32-item

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checklist was used for explicit and comprehensive reporting of the study methods.

Results

Several common themes emerged from patients at different disease stages.

Communication and information

The key needs of patients and carers were structured around 3 areas: context, timing, and content and format of information. Patients and carers described a need to balance honesty and hope, while dealing with a terminal prognosis. They felt that the focus of the consultation at specialist IPF clinics was often disconnected from their experience and the impact of the disease on their everyday life. They also felt that they need to receive information at an appropriate pace triggered by changes in their health status.

Patients and carers showed a good understanding of the overall prognosis for IPF. However, patients had difficulty translating this to their own particular disease progression and to the available management and support options.

Changes in health status

Patients developed their own subjective strategies for monitoring IPF. The findings indicated that patients and carers need different information and that these needs change over time and in particular for those at the progressive stage of the disease. The findings showed that although patients might be reluctant to receive detailed information, the carers need frank information about functional deterioration in order to plan future care.

Functional activity

The risk of social isolation for patients and carers was increased along with patient's health deterioration. Because of the uncertainty about the IPF stages, most patients and carers felt unable to identify potential sources of help and support. Although deteriorating health increased the fears of unemployment and financial status in patients, it extended future emotional and domestic burden on carers.

Understanding of symptoms and medical interventions

Specific concerns were particularly specified from carers in relation to symptom

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monitoring regarding breathing, cough and use of oxygen. Different expectations were observed between patients and carers around oxygen use. Although carers viewed the oxygen use positively as facilitating daily living, patients often perceived it as a failure on their part. Pulmonary rehabilitation was viewed as positive intervention by patients; however, carers felt these strategies were better accepted if offered by a professional rather than by themselves. Patients and carers often compared their situation unfavourably with cancer patients whom they thought to have "help coming from every direction".

Roles and coping strategies of patients and carers

Significant differences were identified in the range of coping strategies employed by patients and carers. Acceptance and adapting to change were the main coping strategies for patients through all stages of the disease. Carers had expressed that they placed the needs of the patient above their own and provided emotional support and motivation. Carers expressed fears about managing their domestic situation in the future and living with feelings of guilt.

Strengths and limitations

Strengths

The aim of the research was clearly specified and the appropriate qualitative methodology was used. The research is a UK-based study that addresses a topic related to NICE guideline CG163 recommendations about information and support. Improvement in psychosocial health and quality of life in patients with IPF was considered as a relevant outcome in the included studies in NICE guideline CG163. That is broadly covered in the current paper.

Limitations

The population for this study was recruited from 2 clinics in the UK, which the service delivery model and setting may have been different from other parts of the UK. In addition none of the patients included in the study received pirfenidone as a part of their treatment. That might have an effect on their experience.

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Impact on guideline

This multicentre, mixed-methods study assessed the care needs of patients with IPF at 4 stages of the disease through semi-structured interviews with patients and paired carers. The study suggests that patients diagnosed with IPF have a clear understanding of their prognosis but little understanding of how their disease will progress and how it will be managed. The evidence indicates that support needed for patients and carers at key transition points. This is in line with NICE guideline CG163 that recommends 'an interstitial lung disease specialist nurse should be available at all stages of the care pathway to provide information and support to people with IPF and their families and carers with the person's consent' (recommendation 1.3.3). Current recommendations promote a supportive approach coordinated by a multidisciplinary team with appropriate skills (recommendations 1.3.1–1.3.4).

NICE has also produced guidance on the components of good patient experience in adult NHS services.

Management – disease-modifying pharmacological interventions

We selected Sun et al. (2016) for a full commentary because it addresses pharmacological management of IPF in relation to guidance on use of oral N-acetylcysteine.

What the guideline recommends

NICE guideline CG163 recommends 'advise the person that oral N-acetylcysteine is used for managing IPF, but its benefits are uncertain' (recommendation 1.5.13).

Methods

The systematic review included studies that compared N-acetylcysteine-treatment group with a control group that reported specified outcomes. Searches only performed on the limited databases (PubMed, EMBASE, Cochrane Central and Google Scholar). No clear search period was specified.

Methodological quality of the included studies was varied and most were of low quality.

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Results

Findings from meta-analysis of 5 trials, with a total of 564 patients, reported no beneficial effect of N-acetylcysteine on changes in forced vital capacity (standard mean difference [SMD]=0.07, 95% confidence interval [CI] 0.13 to 0.27, p=0.52), changes in predicted carbon monoxide diffusing capacity (SMD=0.12, 95% CI 0.06 to 0.30, p=0.18), rates of adverse events (odds ratio [OR]=4.50; 95% CI 0.19 to 106.41, p=0.35), or death rates (OR=1.79, 95% CI 0.3 to 5.12, p=0.28) between the N-acetylcysteine group and the control group. N-acetylcysteine compared with placebo was found to have a significant effect only on decreases in percentage of predicted vital capacity (SMD=0.37, 95% CI 0.13 to 0.62, p=0.003) and decline in 6-minute walking test (SMD=0.25; 95% CI 0.02 to 0.48, p=0.04).

Strengths and limitations

Strengths

The systematic review targeted the exact population and outcomes reported in NICE guideline CG163. All-cause and IPF-related mortality was 1 of the critical outcomes outlined by the guideline that was reported in the study. The study also reported other relevant outcomes including changes in forced vital capacity, changes in predicted carbon monoxide diffusing capacity, and rates of adverse outcomes.

Limitations

No exclusion criteria were specified. Searches were only performed on the limited databases (PubMed, EMBASE, Cochrane Central and Google Scholar). No clear search period was specified. Additional searches were not carried out on the bibliographic databases.

All included studies in the systematic review had risk of biases and generally were assessed as low quality. No sensitivity analysis was performed based on the quality of the studies. It is unclear how and by whom the quality assessment of the studies was performed. The systematic review targeted the exact population and outcomes reported in NICE guideline CG163. However, the stage of the disease was not reported and considered in the analysis of the data.

Studies were heterogeneous in terms of population and interventions. There were

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variations in dosage regimens of N-acetylcysteine in the included studies. There were also variations between the control groups in the included studies. Two of the 5 included studies had no therapy in the control group, whereas 2 had placebo and 1 used bromhexine hydrochloride in the control group. These variations were not taken into account in data analyses in the form of stratified and subgroup analyses, which undermines the validity of the findings, although the random-effects model was used in pooling results of heterogeneous studies.

Impact on guideline

This systematic review evaluated the efficacy of N-acetylcysteine, compared with control, for treating IPF. Findings showed no beneficial effect of N-acetylcysteine on changes in forced vital capacity, changes in predicted carbon monoxide diffusing capacity, rates of adverse events, or death. N-acetylcysteine was found to have a significant effect only on decreases in percentage of predicted vital capacity and decline in 6-minute walking test distance.

NICE guideline CG163 recommends 'advise the person that oral N-acetylcysteine is used for managing IPF, but its benefits are uncertain' (recommendation 1.5.13). The recommendation already acknowledges uncertainty about the benefits of this drug, and no new safety concerns have been raised about its use. Therefore the findings from this study do not impact on current recommendations.

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How we made the decision

We check our guidelines regularly to ensure they remain up to date. We based the decision on surveillance 4 years after the publication of idiopathic pulmonary fibrosis (2013) NICE guideline CG163.

For details of the process and update decisions that are available, see ensuring that published guidelines are current and accurate in developing NICE guidelines: the manual.

Previous surveillance update decisions for the guideline are on our website.

Evidence

We found 24 studies in a search for randomised controlled trials and systematic reviews published between 13 February 2015 and 22 September 2016. We also considered 2 additional studies identified by members of the guideline committee who originally worked on this guideline.

We also considered evidence identified in a previous surveillance 12 years after publication of the guideline.

From all sources, we considered 38 studies to be relevant to the guideline.

We also checked for relevant ongoing research, which will be evaluated again at the next surveillance review of the guideline.

See appendix A: summary of evidence from surveillance and references for all evidence considered.

Views of topic experts

We considered the views of topic experts, including those who helped to develop the guideline.

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Views of stakeholders

Stakeholders commented on the decision not to update the guideline. Overall, 7 stakeholders commented. See appendix B for stakeholders' comments and our responses.

One agreed and 6 disagreed with this proposal. Of the 6 who disagreed, 1 provided comments that were unclear as to why they did not agree with the proposal not to update. Three stakeholders who disagreed with the decision to not update the guideline suggested updating the guideline in accordance with NICE technology appraisal guidance on nintedanib, and asked for clearer statement around the lack of benefit of N-acetylcysteine.

We responded that we will amend the guideline to add a cross-reference to NICE technology appraisal guidance TA379 published in January 2016 from recommendations in section 1.5 in NICE guideline CG163. Concerning N-acetylcysteine, NICE guideline CG163 recommends 'advise the person that oral N-acetylcysteine is used for managing idiopathic pulmonary fibrosis, but its benefits are uncertain' (recommendation 1.5.13). Because the recommendation already acknowledges uncertainty about the benefits of this drug, and no new safety concerns have been raised in review of evidence about its use, there is no impact on current recommendations at this time. This area will be examined again at the next surveillance review of the guideline.

Two stakeholders who also disagreed with the decision to not update the guideline commented that patients' and carers' values and preferences need to be established. In the current surveillance review, none of the new evidence considered was thought to have an impact on the current recommendations on patients' information and support.

NICE has produced guidance on the components of good patient experience in adult NHS services. The related recommendations are in the NICE guideline on patient experience in adult NHS services.

Three stakeholders disagreed with the decision to remove 3 of the research recommendations from the NICE version of the guideline and NICE research database because of relevant ongoing trials; therefore these recommendations will be retained.

See ensuring that published guidelines are current and accurate in developing NICE guidelines: the manual for more details on our consultation processes.

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