Angiotensin-converting enzyme inhibitors (ACE-I) use for the management of paediatric heart failure

1. Which cardiac diseases related to heart failure development do you manage with ACE-I?
Please select all items that apply to you.

☐ Dilated cardiomyopathy
☐ Congenital heart defects
☐ None
☐ Other (please specify in the box below)

[ ]

2. Which paediatric age groups do you treat with ACE-I?
Please select all items that apply to you.

☐ Newborns (0-27 days)
☐ Infants and toddlers (28 days-23 months)
☐ Children (2-11 years)
☐ Adolescents (12-18 years)

3. Which ACE-I do you consider as your first choice for NEWBORNS (0-27 DAYS)?

ACE-I
Please select one item from the list.

☐ Benazepril   ☐ Fosinopril   ☐ Quinapril
☐ Captopril   ☐ Imidapril   ☐ Ramirpil
☐ Cilazapril   ☐ Lisinopril   ☐ Trandolapril
☐ Enalapril   ☐ Moexipril   ☐ Zofenopril
☐ Espirapril   ☐ Perindopril   ☐ Other (please specify in the box below)
                                             [ ]

Starting dose in mg/kg per DOSE
Please type the dose in the box.

[ ]

Target / Maintenance dose in mg/kg per DAY
Please type the dose in the box.

[ ]

In how many doses is the target / maintenance DAILY dose divided?
Please select one item from the list.

☐ One single dose
☐ Two divided doses
☐ Three divided doses
☐ Four divided doses

Figure S2. Questionnaire distributed in the European Survey on the Pharmacological Management of Paediatric Heart Failure. Questionnaire included single-choice, multiple-choice and open questions, and was designed to be completed within 15 minutes. Instructions to facilitate the navigation through the web-questionnaire were provided. Routing filters were implemented; therefore participants were only presented those questions that according to their own answers were applicable to them. Question 3 was displayed according to each age group the participant had selected in question 2.

Why is this ACE-I your first choice for NEWBORNS?
Please select all items that apply to you.

- ☐ More experience with use
- ☐ Most appropriate formulation available
- ☐ More convenient to parents/patients
- ☐ Recommended in guidelines or books
- ☐ Established in hospital protocols
- ☐ No specific reason
- ☐ Other (please specify in the box below)

4. Which ACE-I formulation do you prescribe when the adults formulation is not suitable for a patient?
Please select all items that apply to you.

- ☐ Formulation provided by my hospital pharmacy
- ☐ Formulation provided by community pharmacy
- ☐ Formulation prepared by parents at home using the adults formulation
- ☐ Other (please specify in the box below)

5. What kind of formulation is it?
Please select all items that apply to you.

- ☐ Liquid formulation
- ☐ Capsules
- ☐ Powder
- ☐ Other (please specify in the box below)

6. Do you increase the dose of ACE-I to your target although patient has already improved with a lower dose?
Please select one item.

- ☐ No
- ☐ Yes
- ☐ Sometimes (please specify in the box below)

7. How do you assess the effectiveness of ACE-I in your paediatric patients?
Please select all items that apply to you.

- ☐ Clinical judgement according to changes in signs and symptoms
- ☐ Needs of anticongestive medication
- ☐ Parents' opinion/ perception
- ☐ Clinical scores (e.g. Ross, NYHA)
- ☐ Echocardiographic or radiographic parameters
- ☐ Level of natriuretic peptide
- ☐ Quality of life scores
- ☐ Other (please specify in the box below)

Figure S2. Questionnaire distributed in the European Survey on the Pharmacological Management of Paediatric Heart Failure.

We know, it is difficult to give a simple answer to the following question. Please, we would like you to select the option that most approximates to your practice.

8. If deterioration of the renal function is detected,
which serum creatinine increase relative to baseline value makes you stop increasing the dose of ACE-I?
Please select one item.

☐ 1.1 to 1.4 times
☐ 1.5 to 1.9 times
☐ 2.0 to 2.9 times
☐ 3 times or more
☐ No formal limit used

which serum creatinine increase relative to baseline value makes you withdraw the therapy with ACE-I?
Please select one item.

☐ 1.1 to 1.4 times
☐ 1.5 to 1.9 times
☐ 2.0 to 2.9 times
☐ 3 times or more
☐ No formal limit used

Please add any additional comment that you consider relevant to this question:

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[                                                          ]
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9. If hypotension is detected, do you follow pre-established formal blood pressure limits that make you

	Yes	No
stop increasing the dose of ACE-I? Please select one item.	☐	☐
withdraw the therapy with ACE-I? Please select one item.	☐	☐

10. How do you assess the effectiveness of ACE-I in your paediatric patients?
Please select all items that apply to you.

☐ Percentage decrease relative to baseline value
☐ Absolute blood pressure values according to age
☐ Other (please specify in the box below)

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[                                                          ]
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11. Which of your paediatric patients with congenital heart diseases do you treat with ACE-I?
Please select one option for each congenital heart disease.

	Asymptomatic	Symptomatic	Both	None
Left-to-right shunt lesions	☐	☐	☐	☐
Pressure overloading lesions	☐	☐	☐	☐
Single ventricle lesions	☐	☐	☐	☐
Valve regurgitation	☐	☐	☐	☐

Please add any additional comment that you consider relevant to this question:

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[                                                          ]
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Figure S2. Questionnaire distributed in the European Survey on the Pharmacological Management of Paediatric Heart Failure.

12. Do you use ACE-I for the management of any other congenital heart disease?
Please select one item.

☐ No
☐ Yes (please specify in the box below)

[   ]

13. How long do you treat your patients with congenital heart diseases with ACE-I after heart surgery?
Please select one item.

☐ < 1 month
☐ 1 to 3 months
☐ > 3 months to 6 months
☐ > 6 months
☐ I do not use ACE-I in my patients after heart surgery

Pharmacologic management of paediatric heart failure in patients with dilated cardiomyopathy

14. When treating dilated cardiomyopathy, which drugs do you use as initial therapy of symptomatic patients, who are not dependent on intravenous inotropic/vasoactive drugs (e.g. dobutamine, milrinone, nitroglycerin, levosimendan...)?
Please select all items that apply to you.

☐ Angiotensin-converting enzyme inhibitors (captopril, enalapril...)
☐ Angiotensin receptor blockers (candesartan, losartan...)
☐ Beta-blockers (bisoprolol, carvedilol...)
☐ Loop diuretics (furosemide, torasemide...)
☐ Thiazide diuretics (hydrochlorothiazide...)
☐ Aldosterone antagonist (spironolactone, eplerenone...) ☐ Quality of life scores
☐ Cardiac glycosides (digoxin, digitoxin...)
☐ Other (please specify in the box below):

[   ]

15. Which drug do you add if patients remain symptomatic despite initial therapy?
Please select all items that apply to you.

☐ Angiotensin-converting enzyme inhibitors (captopril, enalapril...)
☐ Angiotensin receptor blockers (candesartan, losartan...)
☐ Beta-blockers (bisoprolol, carvedilol...)
☐ Loop diuretics (furosemide, torasemide...)
☐ Thiazide diuretics (hydrochlorothiazide...)
☐ Aldosterone antagonist (spironolactone, eplerenone...) ☐ Quality of life scores
☐ Cardiac glycosides (digoxin, digitoxin...)
☐ Other (please specify in the box below):

[   ]

16. Do you prescribe drug treatment to asymptomatic patients?
Please select one item.

☐ No
☐ Yes
☐ Sometimes (please specify in the box below)

[   ]

Figure S2. Questionnaire distributed in the European Survey on the Pharmacological Management of Paediatric Heart Failure.

17. Which drug do you use for these asymptomatic patients?
Please select all items that apply to you.

☐ Angiotensin-converting enzyme inhibitors (captopril, enalapril...)
☐ Angiotensin receptor blockers (candesartan, losartan...)
☐ Beta-blockers (bisoprolol, carvedilol...)
☐ Loop diuretics (furosemide, torasemide...)
☐ Thiazide diuretics (hydrochlorothiazide...)
☐ Aldosterone antagonist (spironolactone, eplerenone...) ☐ Quality of life scores
☐ Cardiac glycosides (digoxin, digitoxin...)
☐ Other (please specify in the box below):

[ ]

Feedback and demographic characteristics

18. According to your experience, how would you grade the impact of pharmacological therapy on the course of the disease in your paediatric heart failure patients? (1 no impact, 10 maximal impact)
Please select one item.

☐1 ☐2 ☐3 ☐4 ☐5 ☐6 ☐7 ☐8 ☐9 ☐10

19. Would you like to add any comment that you consider relevant to this survey?
Please select one item.

☐ No
☐ Yes (please specify in the box below)

[ ]

20. How many years of experience do you have caring for children with heart failure?
Please select one item.

☐ < 1 year
☐ 1 to 5 years
☐ > 5 to 10 years
☐ > 10 years

21. In which type of unit do you work?
Please select one item.

☐ Paediatric cardiology
☐ Paediatric critical care
☐ Neonatology
☐ Other (please specify in the box below)

[ ]

22. How many total paediatric beds (not only in your ward) does the hospital you are working in have?
Please type in in the box below.

[ ]

23. In which hospital are you working?
This information will only be used to check how many different hospitals and countries contributed to the results. Please remember that all your answers will be reported anonymously.

Name of the hospital   **City**   **Country**

[ ]   [ ]   [ ]

Figure S2. Questionnaire distributed in the European Survey on the Pharmacological Management of Paediatric Heart Failure.